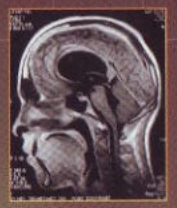
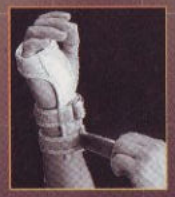
Handbook of
Neuro-Rehabilitation





Amit Agrawal
Dhaval P Shukla

Guest Editor
Prof M Shantharam Shetty

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Handbook of Neuro-Rehabilitation

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Amit Agrawal MS, MCh
Assistant Professor
(Neurosurgery)
BP Koirala Institute of Medical Sciences
Dharan, Nepal

Dhaval P Shukla
Consultant Neurosurgeon
Apollo Hospital
Ahemadabad

Guest Editor
Prof M Shantharam Shetty
Professor and Dean
Department of Orthopaedics
KS Hegde Medical Academy
Deralakatte
Mangalore, Karnataka

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Foreword

The miracles of modern medicine have created scenarios where persons with clinical conditions previously considered "fatal" and/or "permanent" have in fact shown recovery. Inherent in aggressive and advanced medical treatment has also been a phenomenon leading to an increased number of persons in such states relying on life-sustaining treatments. In many cases, such patients remain dependent on these life-sustaining treatments for years. Providing independence and mobility to a person with disability will ultimately improve his/her quality of life. This book will provide basic information to many scientists and clinicians interested in this clinical topic.

The text is designed to be useful for all professionals and particularly students concerned with rehabilitation following neurological disorders. Authors contribute several chapters on a wide array of topics, displaying both dedication and optimism as to the ultimate efficacy of holistic rehabilitation techniques. The book involves a review of basic neurosciences, fundamentals of neurological investigations and clinical assessment and its relevance for acute and postacute treatment. The book covers the dedicated chapters on orthotics and reconstructive surgery. They implore colleagues and practitioners to remain creative and set hopeful goals to help people with nervous system disorders to achieve a new constellation of adaptive capacity following the illness.

Without any reservations, I hope that the book provides a solid foundation to conquer some enigmas in the Neuro-rehabilitation.

Prof CS Agrawal
Acting Rector, Professor and Head
Department of Surgery
BP Koirala Institute of Health Sciences
Dharan, Nepal

Preface

Rehabilitation aims to improve independence and quality of life by maximizing ability and participation. It has been defined by the World Health Organization as "an active process by which those disabled by injury or disease achieve a full recovery, or if a full recovery is not possible, realize their optimal physical, mental and social potential and are integrated into their most appropriate environment." The philosophy of rehabilitation emphasizes patient education and self-management. This philosophy of rehabilitation applies at every stage of the condition, from initial diagnosis to the management of those with severe disability. A successful management strategy must account for the complex pattern of disability that results, together with the possibility that treating one symptom may worsen another. It also is apparent that comprehensive management will invariably require a number of different approaches, including the provision of information, patient education, therapy from a range of disciplines, and drug treatment.

Rehabilitation is an ongoing process in which assessment and therapeutic intervention are reiterative and need a multidisciplinary approach. This book is written by a large number of well-known authors from a variety of disciplines and comprehensively describes the different aspects of the process of neurological rehabilitation. The book is divided into different sections. Each chapter is supported by many pictures, and detailed tables facilitate the comprehensibility of the text.

This book is written for physiotherapy students and professionals to assist in understanding the interrelationship between neurological sciences and rehabilitation sciences. Our attempt is to bridge the gap between medical and physical rehabilitation sciences. This book will provide a broad discussion of neuro-rehabilitation issues, and try to explain basic principles of neurology that can be applied to understanding and solving the problem of patients with physical disability.

Amit Agrawal
Dhaval P Shukla
M Shantharam Shetty

Contributors

- **Prof M Shantharam Shetty**
  Professor and Dean
  Department of Orthopedics
  KS Hegde Medical Academy
  Deralakatte
  Mangalore, Karnataka

- **Dr Amit Agrawal**
  Assistant professor
  Neurosurgery
  BP Koirala Institute of Medical Sciences
  Dharan, Nepal

- **Dr Dhaval P Shukla**
  Consultant Neurosurgeon
  Apollo Hospital
  Ahemadabad

- **Mr Shreekumaran P**
  Professor and Principal
  Nitte Institute of Physiotherapy
  Deralakatte
  Mangalore, Karnataka

- **Mr Dhanesh Kumar KU**
  Associate Professor
  Nitte Institute of Physiotherapy
  Deralakatte
  Mangalore, Karnataka

- **Dr Ranjit Peter**
  Assistant Professor
  Department of ENT
  KS Hegde Medical Academy
  Deralakatte
  Mangalore, Karnataka

- **Dr Sankalp Dwivedi**
  Lecturer in Surgery
  Department of Surgery
  Mahatma Gandhi Institute of Medical Sciences
  Sevagram

- **Dr Naresh Dutt**
  Consultant Physician
  Dhanaura

- **Dr Peter George**
  Assistant Professor
  Department of Medicine
  KS Hegde Medical Academy
  Deralakattes
  Mangalore, Karnataka

- **Lt Col Dr AK Sampath Kumar**
  Associate professor
  Department of Medicine
  KS Hegde Medical Academy
  Deralakatte
  Mangalore, Karnataka

- **Dr Swapnil S Nagvenkar**
  Tutor
  Department of Radiology
  KS Hegde Medical Academy
  Deralakatte
  Mangalore, Karnata...

○ **Dr Vikram Shetty**
Tutor
Department of Orthopedics
KS Hegde Medical Academy
Deralakatte
Mangalore, Karnataka

○ **Dr Gagan Sabharwal**
Senior Resident
Department of Oral and Maxillofacial Surgery
AB Shetty Memorial Institute of Dental Sciences
Deralakatte
Mangalore, Karnataka

○ **Dr Yogaraj KS**
Associate Professor
Department of Anesthesiology
KS Hegde Medical Academy
Deralakatte
Mangalore, Karnataka

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BASIC NEUROSCIENCES

CHAPTER 1

Essential Neuroanatomy

ORGANISATION OF THE NERVOUS SYSTEM

I. Central nervous system
   1. Brain
      a. Forebrain (prosencephalon)
         - Telencephalon (cerebral hemispheres)
         - Diencephalon

      b. Midbrain (mesencephalon)

      c. Hindbrain (rhombencephalon)
         - Medulla oblongata
         - Pons and
         - Cerebellum

   2. Spinal cord

II. Peripheral nervous system
   1. Sensory-somatic nervous system
      a. Cranial nerves—12 pairs
      b. Spinal nerves—31 pairs

   2. Autonomic nervous system
      a. Sympathetic nervous system
      b. Parasympathetic nervous system

BRAIN

Forebrain

Cerebral Hemispheres

Each hemisphere of the cerebrum is subdivided into four lobes (Figure 1.1) visible from the outside:

- Frontal
- Parietal
- Occipital
- Temporal

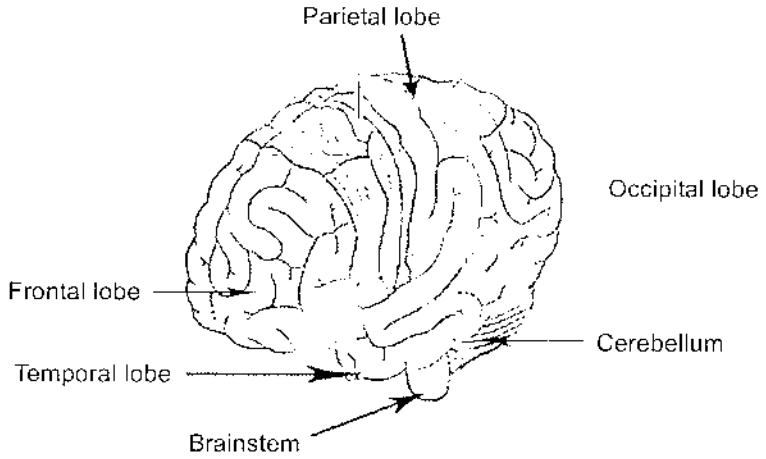

Fig. 1.1 Division and lobes of brain

Frontal Lobe

Front part of the brain; involved in planning, organizing, problem solving, selective attention, personality and a variety of "higher cognitive functions" including behavior and emotions. The frontal lobe is divided from the parietal lobe by the central sulcus.

Prefrontal cortex. The anterior portion of the frontal lobe is called the prefrontal cortex. It is very important for the "higher cognitive functions" and the determination of the personality.

Premotor and motor areas. The posterior of the frontal lobe consists of the premotor and motor areas. Nerve cells that produce movement are located in the motor areas. The premotor areas serve to modify movements.

Occipital Lobe
Region in the back of the brain which processes visual information. Not only is the occipital lobe mainly responsible for visual reception, it also contains association areas that help in the visual recognition of shapes and colors. Damage to this lobe can cause visual deficits.

Parietal Lobe
Parietal lobes of the brain located behind the frontal lobe at the top of the brain. The parietal lobes contain the primary sensory cortex which controls sensation (touch, pressure). Posterior to the primary sensory cortex is a large association area that controls fine sensation (judgment of texture, weight, size, shape).

Right parietal lobe. Damage to this area can cause visuo-spatial deficits (e.g., the patient may have difficulty finding his way around new, or even familiar, places).

Left parietal lobe. Damage to this area may disrupt a patient's ability to understand spoken and/or written language.

Temporal Lobe
There are two temporal lobes, one on each side of the brain located at about the level of the ears. These lobes allow a person to tell one smell from another and one sound from another. They also help in sorting new information and are believed to be responsible for short-term memory.

Right temporal lobe. Mainly involved in visual memory (i.e., memory for pictures and faces).

Left temporal lobe. Mainly involved in verbal memory (i.e., memory for words and names).

The Basal Ganglia

The basal ganglia are a group of anatomically closely related subcortical nuclei. Basal ganglia comprises of five structures on each side of the brain: the caudate nucleus, putamen, and globus pallidus, three large nuclear masses underlying the cortical mantle, and the functionally related subthalamic nucleus (body of Luys) and substantia nigra. The globus pallidus is divided into an external and an internal segment. The substantia nigra is divided into a pars compacta and a pars reticulata. Parts of the thalamus are intimately related to the basal ganglia. The caudate nucleus and the putamen are frequently called the striatum; the putamen and the globus pallidus are sometimes called the lenticular nucleus. The main afferent

connections to the basal ganglia terminate in the striatum. They include the corticostriate projection from all parts of the cerebral cortex. There is also a projection from the centromedian nucleus of the thalamus to the striatum. Damage to these nuclei does not cause weakness, but can cause motor abnormalities.

Movement disorders—chorea, tremors at rest and with initiation of movement, abnormal increase in muscle tone, difficulty initiating movement

Function of Basal Ganglia
Planning and programming of movement
Coverting an abstract thought into voluntary action
They discharge via the thalamus to areas related to the motor cortex, and the corticospinal pathways provide the final common pathway to the motor neurons. In addition, the field potentials in the basal ganglia oscillate, and it has been suggested that the oscillations may have functions like the putative functions of the oscillations of the thalamocortical circuits.
Also play a role in some cognitive processes
Dysarthric form of aphasia

The Diencephalon

The diencephalon consists of the thalamus, hypothalamus, subthalamus, and epithalamus.

The Thalamus
All sensory input (except for olfaction) passes through it on the way up to the somatic-sensory regions of the cerebral cortex and then returns to it from there. Signals from the cerebellum pass through it on the way to the motor areas of the cerebral cortex. Thalamic dysfunction can cause altered level of consciousness, loss of perception and thalamic syndrome (spontaneous pain opposite side of body).

Hypothalamus
Hypothalamus is the seat of the autonomic nervous system. Hypothalamus controls the body temperature, electrolytes and many other important functions.

Midbrain

The midbrain serves as the nerve pathway of the cerebral hemispheres and contains auditory and visual reflex centers. The midbrain along with the medulla and pons are often referred to as the "brainstem".

Brainstem

This is the lower extension of the brain where it connects to the spinal cord. Neurological functions located in the brainstem include those necessary for survival (breathing, digestion, heart rate, blood pressure) and for arousal (being awake and alert). Most of the cranial nerves come from the brainstem. The brainstem is the pathway for all fiber tracts passing up and down from peripheral nerves and spinal cord to the highest parts of the brain (Table 1.1).

Table 1.1 Brainstem and location of cranial nerve nuclei

Level	Nuclei
Midbrain	III, IV, mesencephalic V
Pons	V (main nucleus)
Caudal pons	VI, VII
Ponto-medullary junction	VIII
Medulla	N. of the descending tract of V. N. ambiguus N. tractus solitarius Motor X, XII
Cervical cord	XI

Hindbrain

The main structures of the hindbrain (rhombencephalon) are:

- Medulla oblongata
- Pons
- Cerebellum

Medulla Oblongata

The medulla oblongata functions primarily as a relay station for the crossing of motor tracts between the spinal cord and the brain. It also contains the respiratory, vasomotor and cardiac centers, as well as many mechanisms for controlling reflex activities such as coughing, gagging, swallowing and vomiting.

Pons

The pons is a bridge-like structure which links different parts of the brain and serves as a relay station from the medulla to the higher cortical

structures of the brain. Nerve impulses coming from the eyes, ears, and touch receptors are sent on to the cerebellum. The pons also participates in the reflexes that regulate breathing.

Cerebellum (Table 1.2)

The cerebellum sits astride the main sensory and motor systems in the brainstem. It is connected to the brainstem on each side by a superior peduncle (brachium conjunctivum), middle peduncle (brachium pontis), and inferior peduncle (restiform body). The medial vermis and lateral cerebellar hemispheres are more extensively folded and fissured than the cerebral cortex; the cerebellum weighs only 10% as much as the cerebral cortex, but its surface area is about 75% of that of the cerebral cortex.

Anatomically, the cerebellum is divided into three parts by two transverse fissures. The posterolateral fissure separates the medial nodulus and the lateral flocculus on either side from the rest of the cerebellum, and the primary fissure divides the remainder into an anterior and a posterior lobe. Lesser fissures divide the vermis into smaller sections (ten primary lobules numbered I-X from superior to inferior).

Table 1.2 Functional divisions of the cerebellum

Vestibulo-cerebellum	Phylogenetically the oldest part	Vestibular connections	Equilibrium and learning-induced changes in the VOR
Spinocere-bellum		Receives proprioceptive input from the body	Smoothes and coordinates movements
Neocerebellum	Newest	Interact with the motor cortex	Planning and programming movements

SPINAL CORD

As the injury to different regions of the spinal cord will produce predictable patterns, a knowledge of spinal cord anatomy is important. The spinal cord is divided into 31 segments, each with a pair of anterior (motor) and dorsal (sensory) spinal nerve roots. On each side, the anterior and dorsal nerve roots combine to form the spinal nerve as it exits from the vertebral column through the neuroforamina. The spinal cord extends from the base of the skull and terminates near the lower margin of the L1 vertebral body.

Thereafter, the spinal canal contains the lumbar, sacral, and coccygeal spinal nerves that comprise the cauda equina.

Spinal injuries proximal to L1, above the termination of the spinal cord, often involve a combination of spinal cord lesions and segmental root or spinal nerve injuries. The spinal cord itself is organized into a series of tracts or neuropathways that carry motor (descending) and sensory (ascending) information. These tracts are organized anatomically within the spinal cord. The corticospinal tracts are descending motor pathways located anteriorly within the spinal cord. Axons extend from the cerebral cortex in the brain as far as the corresponding segment, where they form synapses with motor neurons in the anterior (ventral) horn. They decussate (cross over) in the medulla prior to entering the spinal cord. The dorsal columns are ascending sensory tracts that transmit light touch, proprioception, and vibration information to the sensory cortex. They do not decussate until they reach the medulla. The lateral spinothalamic tracts transmit pain and temperature sensation. These tracts usually decussate within 3 segments of their origin as they ascend. The anterior spinothalamic tract transmits light touch. Autonomic function traverses within the anterior interomedial tract. Sympathetic nervous system fibers exit the spinal cord between C7 and L1, while parasympathetic system pathways exit between S2 and S4. Injury to the corticospinal tract or dorsal columns, respectively, results in ipsilateral paralysis or loss of sensation of light touch, proprioception, and vibration. Unlike injuries of the other tracts, injury to the lateral spinothalamic tract causes contralateral loss of pain and temperature sensation. Because the anterior spinothalamic tract also transmits light touch information, injury to the dorsal columns may result in complete loss of vibration sensation and proprioception but only partial loss of light touch sensation.

Autonomic Pathways

Autonomic function is transmitted in the anterior interomedial tract. The sympathetic nervous system fibers exit from the spinal cord between C7 and L1. The parasympathetic system nerves exit between S2 and S4. Therefore progressively higher spinal cord lesions or injury causes increasing degrees of autonomic dysfunction. Neurogenic shock is characterized by severe autonomic dysfunction, resulting in hypotension, relative bradycardia, peripheral vasodilation, and hypothermia. It does not usually occur with SCI below the level of T6. Shock associated with an SCI involving the lower thoracic cord must be considered haemorrhagic until proven otherwise.

Blood Supply

The blood supply of the spinal cord consists of 1 anterior and 2 posterior spinal arteries. The anterior spinal artery supplies the anterior two thirds of the cord. Ischaemic injury to this vessel results in dysfunction of the corticospinal, lateral spinothalamic, and autonomic interomedial pathways. Anterior spinal artery syndrome involves paraplegia, loss of pain and temperature sensation, and autonomic dysfunction. The posterior spinal arteries primarily supply the dorsal columns. The anterior and posterior spinal arteries arise from the vertebral arteries in the neck and descend from the base of the skull. Various radicular arteries branch off the thoracic and abdominal aorta to provide collateral flow. The primary watershed area of the spinal cord is the midthoracic region. Because of its peculiar blood supply vascular injury may cause a cord lesion at a level several segments higher than the level of spinal injury. For example, a lower cervical spine fracture may result in disruption of the vertebral artery that ascends through the affected vertebra resulting in ischaemic injury to high cervical cord. At any given level of the spinal cord, the central part is a watershed area. Cervical hyperextension injuries may cause ischaemic injury to the central part of the cord, causing a central cord syndrome.

PERIPHERAL NERVOUS SYSTEM

Sensory-somatic Nervous System

a. **Cranial nerves**—12 pairs (Table 1.3)
b. **Spinal nerves**—31 pairs

Sensory neurons running from stimulus **receptors** that inform the CNS of the stimuli.

Motor neurons running from the CNS to the **muscles and glands**—called **effectors**—that take action.

The Spinal Nerves

All of the spinal nerves are "mixed"; that is, they contain both sensory and motor neurons.

- Cervical—8
- Thoracic—12
- Lumbar—5
- Sacral—5
- Coccygeal—1

Table 1.3 Summary of cranial nerves

Nerves	Type	Function
I Olfactory	Sensory	Olfaction (smell)
II Optic	Sensory	Vision
III Oculomotor	Motor	Eyelid and eyeball muscles
IV Trochlear	Motor	Eyeball muscles
V Trigeminal	Mixed	Sensory: facial and mouth sensation Motor: chewing
VI Abducens	Motor	Eyeball movement
VII Facial	Mixed	Sensory: taste Motor: facial muscles and salivary glands
VIII Auditory	Sensory	Hearing and balance
IX Glossopharyngeal	Mixed	Sensory: taste Motor: swallowing
X Vagus	Mixed	Main nerve of the parasympathetic nervous system (PNS)
XI Accessory	Motor	Swallowing; moving head and shoulder
XII Hypoglossal	Motor	Tongue muscles

Autonomic Nervous System

The autonomic nervous system consists of sensory neurons and motor neurons that run between the central nervous system (especially the hypothalamus and medulla oblongata) and various internal organs (heart, lungs, viscera etc.)

Preganglionic neurons arise in the CNS and run to a ganglion in the body. Here they synapse with ***postganglionic*** neurons, which run to the effector organ (cardiac muscle, smooth muscle, or a gland).

Sub-divisions of the autonomic nervous system

- Sympathetic nervous system
- Parasympathetic nervous system

The Sympathetic Nervous System

The preganglionic motor neurons of the sympathetic system arise in the spinal cord. They pass into sympathetic ganglia which are organized into two chains that run parallel to and on either side of the spinal cord.

The preganglionic neuron may do one of three things in the sympathetic ganglion:

- Synapse with postganglionic neurons which then reenter the spinal nerve and ultimately pass out to the sweat glands and the walls of blood vessels near the surface of the body.
- Pass up or down the sympathetic chain and finally synapse with post-ganglionic neurons in a higher or lower ganglion
- Leave the ganglion by way of a cord leading to special ganglia (e.g., the solar plexus) in the viscera. Here it may synapse with postganglionic sympathetic neurons running to the smooth muscular walls of the viscera.

However, some of these preganglionic neurons pass right on through this second ganglion and into the adrenal medulla. Here they synapse with the highly-modified postganglionic cells that make up the secretory portion of the adrenal medulla.

The neurotransmitter of the preganglionic sympathetic neurons is acetylcholine (ACh). It stimulates action potentials in the postganglionic neurons.

The neurotransmitter released by the postganglionic neurons is noradrenaline (also called norepinephrine).

The Parasympathetic Nervous System

The main nerves of the parasympathetic system are the tenth cranial nerves, the vagus nerves. They originate in the medulla oblongata. Other preganglionic parasympathetic neurons also extend from the brain as well as from the sacral spinal cord (S2, 3, 4).

Each preganglionic parasympathetic neuron synapses with just a few postganglionic neurons, which are located near – or in – the effector organ,

a muscle or gland. Acetylcholine (ACh) is the neurotransmitter at all the pre- and many of the postganglionic neurons of the parasympathetic system.

Functions of Autonomic Nervous System (Table 1.4)

It is responsible for monitoring conditions in the internal environment and bringing about appropriate changes in them. The contraction of both smooth muscle and cardiac muscle is controlled by motor neurons of the autonomic system.

The actions of the autonomic nervous system are largely involuntary (in contrast to those of the sensory-somatic system). It also differs from the sensory-somatic system in using two groups of motor neurons to stimulate the effectors instead of one.

Table 1.4 Functions of ANS

Organ	Parasympathetic action	Sympathetic action
Pupils	Constriction	Dilatation
Salivary glands	Stimulates salivation	Inhibits salivation
Heart	Slows heart beat	Accelerates heart beat
Bronchi	Constriction	Dilatation
Stomach and small intestine	Stimulates peristalsis and secretion	Inhibits peristalsis and secretion
Liver and gall bladder	Stimulates release of bile	Conversion of glycogen to glucose
Adrenal glands	—	Secretion of adrenaline and noradrenaline
Urinary bladder	Contraction	Inhibition of contraction

CHAPTER 2

Essential Neurophysiology

Neurons are the basic building blocks of the nervous system and transmit the nerve impulses (Figure 2.1). A cortical neuron may receive impulses from tens or even hundreds of thousands of neurons.

NERVE CELLS

The nerve cell may be divided on the basis of its structure and function into three main parts:

Cell Body or Soma

The body of a nerve cell is similar to that of all other cells. The cell body includes the nucleus, mitochondria, endoplasmic reticulum, ribosomes, and other organelles.

Dendrites

The short processes of the cell body, the dendrites, receive impulses from other cells and transfer them to the cell body (afferent signals). These dendrites have small knobby projections called dendritic spines. These knobs are also called terminal buttons or axon telodendria. They contain granules or vesicles in which the synaptic transmitters secreted by the nerves are stored.

Axons

The long nerve fiber, the axon, transfers the signal from the cell body to another nerve or to a muscle cell. Mammalian axons are usually about 1–20 μm in diameter. Axon originates from a somewhat thickened area of the cell body, the axon hillock. The first portion of the axon is called the initial segment. The axon divides into terminal branches, each ending in a number

of synaptic knobs. The axon may be covered with an insulating layer called the myelin sheath, which is formed by Schwann cells. The myelin sheath is not continuous but divided into sections, separated at regular intervals by the nodes of Ranvier (Figure 2.2).

CLASSIFICATION OF NERVE FIBERS

Group A Fibers

Somatic sensory and motor fibers serving skin, muscles, joints; largest diameter; thick myelin sheaths (15–150 m/sec).

Group B Fibers

Lightly myelinated, intermediate diameter (3–15 m/sec)

Group C Fibers

Smallest diameter, unmyelinated (1 m/sec)

Myelinated Nerve Fibres

The myelinated axons are wrapped by a sheath of myelin (a protein-lipid complex). Outside the CNS, the myelin is produced by Schwann cells, glia-like cells found along the axon. The myelin sheath envelops the axon except at its ending and at the nodes of Ranvier.

Non-myelinated Nerve Fibres

These are simply surrounded by Schwann cells without the wrapping of the Schwann cell membrane around the axon that produces myelin.

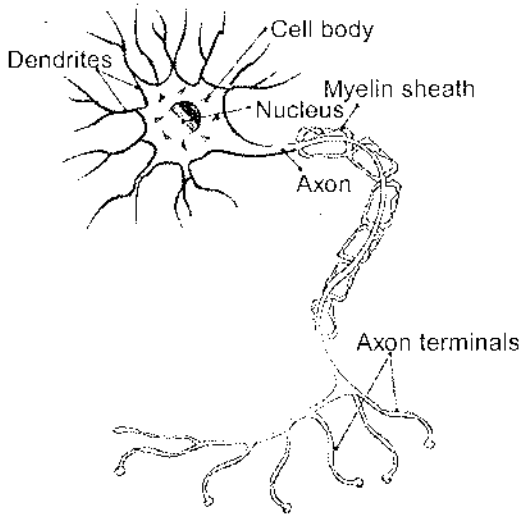

Fig. 2.1 The major components of a neuron

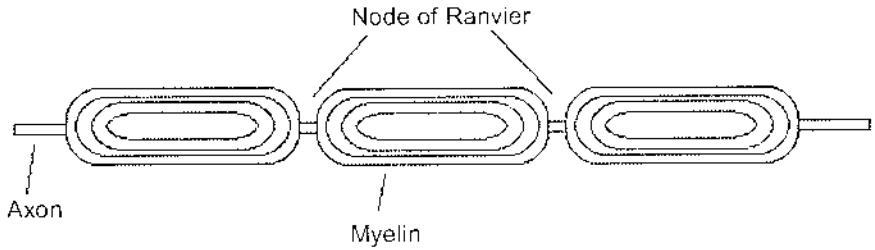

Fig. 2.2 Node of Ranvier

THE SYNAPSE (FIGURE 2.3)

The junction between an axon and the next cell with which it communicates is called the synapse. Information proceeds from the cell body unidirectionally over the synapse, first along the axon and then across the synapse to the next nerve or muscle cell. The part of the synapse that is on the side of the axon is called the presynaptic terminal; that part on the side of the adjacent cell is called the postsynaptic terminal. Between these terminals, there exists a gap, the synaptic cleft, with a thickness of 10–50 nm. The fact that the impulse transfers across the synapse only in one direction, from the presynaptic terminal to the postsynaptic terminal, is due to the release of a chemical transmitter by the presynaptic cell. The synapse between a motor nerve and the muscle it innervates is called the neuromuscular junction.

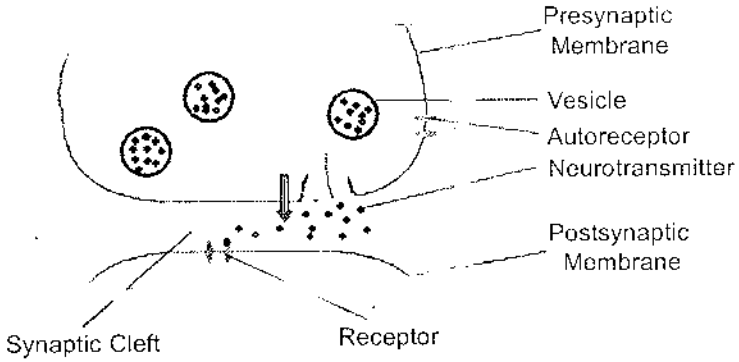

Fig. 2.3 Simplified illustration of the anatomy of the synapse

Types of Synapses

Electrical Synapse

Relatively uncommon; a gap junction between two neurons, allows ions to move directly from one neuron to the next; thus neurons are electrically coupled.

Found in regions of the brain responsible for stereotyped movements, rapid movements of eyes; also common embryonically.

Chemical Synapses

Specialized for release and reception of chemical neurotransmitters (NT); NTs function to open/close chemically-gated channels that influence membrane permeability and hence membrane potential.

Chemical synapse has two parts: a knob-like axon terminal full of synaptic vesicles containing NT molecules; and a region of membrane of postsynaptic neuron that contains receptors.

CLASSIFICATION OF NEURONS

Structural Classification: Based on Number of Processes Extending from Soma

Multipolar Neuron (Figure 2.4a)
Multipolar neurons are so-named because they have many (multi-) processes that extend from the cell body: lots of dendrites plus a single axon. Functionally, these neurons are either motor (conducting impulses that will cause activity such as the contraction of muscles) or association (conducting impulses and permitting 'communication' between neurons within the central nervous system).

Unipolar Neuron (Figure 2.4b)
Unipolar neurons have but one process from the cell body. However, that single, very short, process splits into longer processes (a dendrite plus an axon). Unipolar neurons are sensory neurons – conducting impulses into the central nervous system.

Bipolar Neuron (Figure 2.4c)
Bipolar neurons have two processes – one axon and one dendrite. These neurons are also sensory. For example, bipolar neurons can be found in the retina of the eye.

Functional Classification: According to Direction of Impulse Propagation Relative to CNS

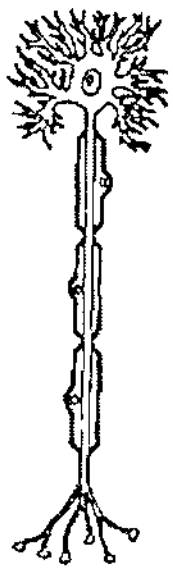

Fig. 2.4a Multipolar neuron

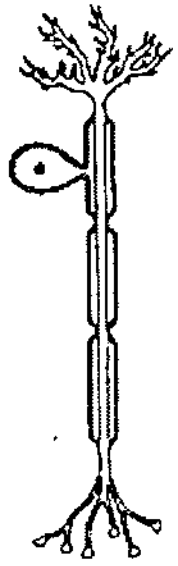

Fig. 2.4b Unipolar neuron

Fig. 2.4c Bipolar neuron

Sensory (afferent) Neurons
Conduct sensory input towards CNS; mostly unipolar; cell bodies in sensory ganglia outside CNS.

Motor (efferent) Neurons
Conduct nerve impulses away from CNS to effector organs; multipolar; cell bodies in CNS (except some ANS), axons bunched as nerves.

Interneurons (association)
Lie between sensory and motor neurons in neural pathways; multipolar, usually confined to CNS.

EXCITABILITY OF NERVE CELL

If a nerve cell is stimulated, the transmembrane voltage necessarily changes. The stimulus may be electrical, chemical, or mechanical. The stimulation may be excitatory (i.e., depolarizing; characterized by a change of the potential inside the cell relative to the outside in the positive direction, and hence by a decrease in the normally negative resting voltage) or inhibitory (i.e., hyperpolarizing, characterized by a change in the potential inside the cell relative to the outside in the negative direction, and hence by an increase in the magnitude of the membrane voltage).

Resting Membrane Potential

Resting membrane potential is due to unequal distribution of ions on the two sides of the nerve cell membrane. This generally measures about 70 millivolts (with the inside of the membrane negative with respect to the outside). Resting membrane potential is expressed as -70 mV, and the minus means that the inside is negative relative to (or compared to) the outside.

Action Potential

An action potential is a very rapid change in membrane potential that occurs when a nerve cell membrane is stimulated. Specifically, the membrane potential goes from the resting potential (typically -70 mV) to some positive value (typically about +30 mV) in a very short period of time (just a few milliseconds).

The stimulus causes the sodium channels to open and, sodium then diffuses rapidly into the nerve cell. All these positively-charged sodiums rushing in causes the membrane potential to become positive (the inside of the membrane is now positive relative to the outside). The sodium channels open only briefly, then close again.

The potassium channels then open, and, because there is more potassium inside the membrane than outside, positively-charged potassium ions diffuse out. As these positive ions go out, the inside of the membrane once again becomes negative with respect to the outside.

Threshold Stimulus & Potential

Action potentials occur only when the membrane is stimulated (depolarized) enough so that sodium channels open completely. The minimum stimulus needed to achieve an action potential is called the threshold stimulus.

The threshold stimulus causes the membrane potential to become less negative (because a stimulus, no matter how small, causes a few sodium channels to open and allows some positively-charged sodium ions to diffuse in).

If the membrane potential reaches the threshold potential (generally 5-15 mV less negative than the resting potential), the voltage-regulated sodium channels all open. Sodium ions rapidly diffuse inward, and depolarization occurs.

All-or-None Law

All-or-None Law – action potentials occur maximally or not at all. In other words, there's no such thing as a partial or weak action potential. Either the threshold potential is reached and an action potential occurs, or it isn't reached and no action potential occurs.

Refractory periods

Absolute Refractory Period

During an action potential, a second stimulus will not produce a second action potential (no matter how strong that stimulus is).

Corresponds to the period when the sodium channels are open (typically just a millisecond or less).

Relative Refractory Periods

Another action potential can be produced, but only if the stimulus is greater than the threshold stimulus.

Corresponds to the period when the potassium channels are open (several milliseconds).

The nerve cell membrane becomes progressively more 'sensitive' (easier to stimulate) as the relative refractory period proceeds. So, it takes a very strong stimulus to cause an action potential at the beginning of the relative refractory period, but only slightly above threshold stimulus to cause an action potential near the end of the relative refractory period.

IMPULSE CONDUCTION

An impulse is simply the movement of action potentials along a nerve cell. Action potentials are localized (only affect a small area of nerve cell membrane). So, when one occurs, only a small area of membrane depolarizes (or 'reverses' potential). As a result, for a split second, areas of membrane adjacent to each other have opposite charges (the depolarized

membrane is negative on the outside and positive on the inside, while the adjacent areas are still positive on the outside and negative on the inside). An electrical circuit (or 'mini-circuit') develops between these oppositely-charged areas (or, in other words, electrons flow between these areas). This 'mini-circuit' stimulates the adjacent area and, therefore, an action potential occurs. This process repeats itself and action potentials move down the nerve cell membrane. This 'movement' of action potentials is called an impulse.

Conduction Velocity

Impulses typically travel along neurons at a speed of anywhere from 1 to 120 meters per second.

The speed of conduction can be influenced by:

- The diameter of a fiber
- Temperature
- The presence or absence of myelin
  Neurons with myelin (or myelinated neurons) conduct impulses much faster than those without myelin.

Saltatory Conduction

Between areas of myelin are non-myelinated areas called the nodes of Ranvier. Because fat (myelin) acts as an insulator, membrane coated with myelin will not conduct an impulse. So, in a myelinated neuron, action potentials only occur along the nodes and, therefore, impulses 'jump' over the areas of myelin – going from node to node in a process called saltatory conduction (with the word saltatory meaning 'jumping'):

Because the impulse 'jumps' over areas of myelin, an impulse travels much faster along a myelinated neuron than along a non-myelinated neuron.

Types of neurotransmitters

Excitatory Neurotransmitters

Excitatory neurotransmitters are those that make membrane potential less negative (via increased permeability of the membrane to sodium) and therefore, tend to 'excite' or stimulate the postsynaptic membrane (e.g., acetylcholine).

Inhibitory Neurotransmitters

Inhibitory neurotransmitters are those that make membrane potential more negative (via increased permeability of the membrane to potassium) and therefore, tend to 'inhibit' (or make less likely) the transmission of an impulse. Examples of inhibitory neurotransmitters include gamma aminobutyric acid (GABA) and beta-endorphin.

Summation

- Temporal summation – transmission of an impulse by rapid stimulation of one or more pre-synaptic neurons
- Spatial summation – transmission of an impulse by simultaneous or nearly simultaneous stimulation of two or more presynaptic neurons.

SUPPORTING CELLS

CNS supporting Cells (Table 2.1)

Neuroglia, branching processes with central body, outnumber neurons 9:1.

- Astrocytes: star shaped, most common; radial projections cling neurons to capillaries, role in making exchanges between neurons and capillaries.
- Microglia: the macrophages of the CNS
- Ependymal cells: range from squamous to columnar, line central cavities of brain/spinal cord
- Oligodendrocytes; wrap themselves around thicker neuron fibers of CNS forming insulating covering, myelin sheath.

PNS supporting Cells

- Satellite cells: surround neuron cell bodies in ganglia, role in chemical environment control.
- Schwann cells: wrap themselves around thicker nerve fibers, form myelin sheaths.

Table 2.1 Types of glial cells and their function

Type of cell	Function
Microglia	Scavenger cells that resemble tissue macrophages
Oligodendrogliocytes	Involved in myelin formation
Astrocytes	Fibrous astrocytes contain intermediate filaments, found primarily in white matter Protoplasmic astrocytes are found in gray matter and have granular cytoplasm

REFLEXES

Reflex Arc

Reflex arc is the basic unit of integrated reflex activity and it consists of a sense organ, an afferent neuron, one or more synapses in a central integrating station or sympathetic ganglion, an efferent neuron, and an

effector. In reflex arc afferent neurons enter via the dorsal roots or cranial nerves and have their cell bodies in the dorsal root ganglia or in the ganglia on the cranial nerves. The efferent fibers leave via the ventral roots or corresponding motor cranial nerves.

Reflex Pathway (Figure 2.5)

Activity in the reflex arc starts in a sensory receptor with a receptor potential whose magnitude is proportionate to the strength of the stimulus. This generates all-or-none action potentials in the afferent nerve, the number of action potentials being proportionate to the size of the generator potential. All-or-none responses are generated in the efferent nerve to produce action potentials that bring about muscle contraction.

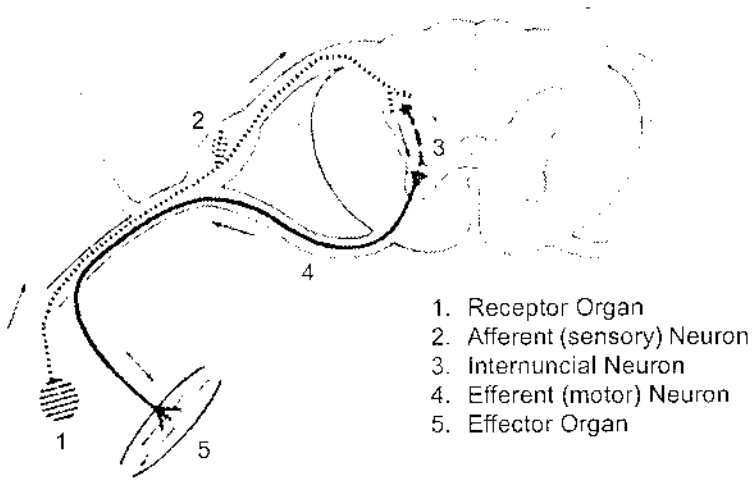

Fig. 2.5 Reflex pathway

Types of Reflexes

Monosynaptic Reflexes

The simplest reflex arc is one with a single synapse between the afferent and efferent neurons. Such arcs are monosynaptic, and reflexes occurring in them are monosynaptic reflexes.

The Stretch Reflex

When a skeletal muscle with an intact nerve supply is stretched, it contracts. This response is called the stretch reflex. Stretch reflexes are the best known and studied monosynaptic reflexes in the body. Clinical examples of monosynaptic stretch reflex include knee jerk, triceps jerk and ankle jerk.

Reciprocal Innervation

When a stretch reflex occurs, the muscles that antagonize the action of the muscle involved (antagonists) relax.

Inverse Stretch Reflex

Up to a point, the harder a muscle is stretched, the stronger is the reflex contraction. However, when the tension becomes great enough, contraction suddenly ceases and the muscle relaxes. This relaxation in response to strong stretch is called the inverse stretch reflex or autogenic inhibition.

Polysynaptic Reflexes

Reflex arcs in which one or more interneurons are interposed between the afferent and efferent neurons are polysynaptic.

Withdrawal Reflex

The withdrawal reflex is a typical polysynaptic reflex that occurs in response to a noxious and usually painful stimulation of the skin or subcutaneous tissues and muscle. The response is flexor muscle contraction and inhibition of extensor muscles, so that the part stimulated is flexed and withdrawn from the stimulus.

NEURONAL PLASTICITY

Synaptic plasticity is a property of adult as well as developing or young cortex, and reflects how synaptic strength changes with experience. Human functional neuroimaging studies have demonstrated changes in neural activity patterns as a behaviour or a response is learned. In nonhuman primates, this can be investigated by recording the firing pattern of cortical neurons.

Physiological Basis

How can neuronal firing patterns lead to a remodeled cortex?

Short-term changes, or memory, result from the strengthening of existing synapses. This occurs via the covalent modification of existing proteins after the activation of second-messenger systems.

Long-term memory requires neuronal gene expression and protein synthesis, resulting in the growth of new synaptic connections.

Role of Neurotransmitters

Neurotransmitters modulate the changes associated with learning and synaptic strengthening:

- Dopamine
- Acetylcholine and norepinephrine
- Serotonin
- Intracellular calcium
- Nitric oxide
- Protein kinases (enzymes that phosphorylate multiple substrates, altering their functioning, altering neuronal physiology, and increasing

CHAPTER 3

Neurological Examination

A thorough neurological history allows the clinician to define the patient problem and, along with the physical examination, assists in formulating an aetiologic/pathologic diagnosis in most cases. Neurological examination begins with the gathering of an accurate patient history and information about the course of the present injury. A sytematic and careul neurological examination is mandatory for the management of patients with neurological disorders (Box 3.1, 3.2; Table 3.1). To perform this one also needs some basic instruments (Box 3.3). All clinical details and findings should be carefully noted and analyzed at the end of examination to make a diagnosis and to plan out a management protocol for the patient.

Box 3.1 Systematic approach to a patient

Higher mental functions
Cranial nerves
Motor
Coordination and gait
Reflexes
Sensory special tests

Box 3.2 Mental status examination

Orientation
Time (date, day, season, year, month)
Place (country, state/province, town, hospital, ward)
Person
Language
Speech (dysphasias, dysarthria, dysphonia)
Pt repeats statement
Pt follows command (Take paper in right hand, fold in half, put it on table)
Registration
Attention and calculation
Recall
Memory
Immediate, recent, remote

Box 3.3 Equipment necessary for neurological examination

Reflex hammer
128 and 512 (or 1024) Hz tuning forks
A Snellen eye chart or pocket vision card
Pen light
Otoscope
Wooden handled cotton swabs
Paper clips
Ophthalmoscope

Table 3.1 Glasgow Coma scale

Eye opening	4-Spontaneous 3-To command 2-To pain 1-None
Verbal response	5-Oriented 4-Confused conversation 3-Inappropriate words 2-Incomprehensible 1-None
Motor response	6-Obeys command 5-Localizes pain 4-Withdraws from pain 3-Abnormal flexion to pain 2-Extension response to pain 1-None

CRANIAL NERVES

I - Olfactory

Ask the patient about smell
Occlude each nostril sequentially. Use a mild test stimulus such as soap, toothpaste, coffee, or lemon oil. With the eyes closed, the patient sniffs and tries to identify the stimulus.

II - Optic

Optic Fundi

Examine with ophthalmoscope

Visual Acuity

- Position the patient 20 feet in front of the Snellen eye chart
- Have the patient cover one eye at a time with a card.
- Ask the patient to read progressively smaller letters until he can go no further.
- Record the smallest line the patient read successfully.
- Repeat with the other eye.

Visual Fields (By confrontation)

- Stand two feet in front of the patient and ask the patient to look into your eyes.
- Hold your hands about one foot away from the patient's ears, and wiggle a finger on one hand.
- Ask the patient to indicate which side they see the finger move.
- Repeat two or three times to test both temporal fields.

Pupillary Reactions to Light

- Dim the room lights as necessary.
- Ask the patient to look into the distance.
- Shine a bright light obliquely into each pupil in turn.
- Look for both the direct (same eye) and consensual (other eye) reactions.
- Record pupil size in mm and any asymmetry or irregularity.

Pupillary Reactions to Accommodation

- Hold your finger about 10 cm from the patient's nose.
- Ask them to alternate looking into the distance and at your finger.
- Observe the pupillary response in each eye.

III, IV and VII - Oculomotor, Trochlear, and Abducens

- Observe for any ptosis

Extraocular Movements

- Stand or sit 3 to 6 feet in front of the patient.
- Ask the patient to follow your finger with his eyes without moving his head.
- Check gaze in the six cardinal directions using a cross or "H" pattern.
- Check for nystagmus.
- Check convergence by moving your finger toward the bridge of the patient's nose

V - Trigeminal

Muscles of Mastication

- Ask patient to both open his mouth and clench his teeth.
- Palpate the temporal and massetter muscles as he does this.

Pain Sensation (Check in all three divisions)

- Use a suitable sharp object to test the forehead, cheeks, and jaw on both sides.
- Substitute a blunt object occasionally and ask the patient to report "sharp" or "dull."

Temperature

- Test the three divisions for temperature sensation with a tuning fork heated or cooled by water.
- Test the three divisions for sensation to light touch using a wisp of cotton.

Corneal Reflex

- Ask the patient to look up and away.
- From the other side, touch the cornea lightly with a fine wisp of cotton.
- Look for the normal blink reaction of both eyes.
- Repeat on the other side.

VII - Facial

Ask patient to do the following and look for any facial asymmetry, lag, weakness, or asymmetry:

- Raise eyebrows
- Close both eyes to resistance
- Smile
- Frown
- Show teeth
- Puff out cheeks

VIII - Acoustic

Hearing

Weber's Test

- Helps in lateralization
- Use a 512 Hz or 1024 Hz tuning fork.
- Start the fork vibrating by tapping it on your opposite hand.
- Place the base of the tuning fork firmly on top of the patient's head.
- Ask the patient where the sound appears to be coming from (normally in the midline).

Rinne's Test

- Compares air and bone conduction
- Use a 512 Hz or 1024 Hz tuning fork.
- Start the fork vibrating by tapping it on your opposite hand.
- Place the base of the tuning fork against the mastoid bone behind the ear.
- When the patient no longer hears the sound, hold the end of the fork near the patient's ear (air conduction is normally greater than bone conduction).

IX and X- Glossopharyngeal and Vagus Nerve

Patient's voice—hoarse or nasal
Ask patient to swallow
Ask patient to say "Ah"
Watch the movements of the soft palate and the pharynx.

Gag Reflex

Stimulate the back of the throat on each side.
Watch for movements of posterior pharyngeal wall

XI - Accessory

Look for atrophy or asymmetry of the trapezius muscles.
Ask patient to shrug shoulders against resistance and compare the strength on either side.
Ask patient to turn his head against resistance. Watch and palpate the sternomastoid muscle on the opposite side.

XII - Hypoglossal

Observe the tongue as it lies in the mouth
Listen to the articulation of the patient's words.
Ask patient to:

- Protrude tongue
- Move tongue from side to side

MOTOR

Observation
Involuntary movements
Muscle symmetry
- Left to Right
- Proximal vs. Distal

Atrophy (Hands, shoulders, and thighs)
Gait

Muscle Tone

There is normally a small, continuous resistance to passive movement.
- Flex and extend the patient's fingers, wrist, and elbow.
- Flex and extend patient's ankle and knee.
- Observe for decreased (flaccid) or increased (rigid/spastic) tone.

Muscle Strength

Test strength by having the patient move against your resistance and grade strength (Box 3.4). Always compare one side to the other.

Box 3.4

0-No movement
1-Flicker
2-Movement, but not against gravity
3-Movement against gravity, but not resistance
4-Movement against resistance, but not entirely normal
5-Normal

Table 3.2 Rapid assessment of level of nerve root injury

Root value	Major function
C4	Spontaneous breathing
C5	Shoulder shrug, deltoid
C6	Biceps, wrist extension
C7	Triceps, wrist flexion
C8/T1	Finger flexion
T1–12	Intercostals, abdominals
L1/L2	Hip flexion
L2/L3/L4	Hip adduction, quadriceps
L4/L5	Hip abduction
L5	Great toe dorsiflexion
S1/S2	Foot plantar flexion
S2–S4	Rectal tone

Table 3.3 Joint movements and motor root value

Joint	Movement (root value)
Shoulder	Abduction (C5) Adduction (C5–C7)
Elbow	Flexion (C5–C6) Extension (C7)
Wrist	Flexion (C7–8) Extension (C7)
Finger	Flexion (C7–8) Extension (C7) Abduction (T1)
Hip	Flexion (L1/L2) Extension (L5/S1)
Knee	Flexion (S1) Extension (L3/L4)
Ankle	Dorsiflexion (L4) Plantar flexion (S1/S2)

Pronator Drift

Ask the patient to stand for 20–30 seconds with both arms straight forward, palms up, and eyes closed.

Instruct the patient to keep the arms still while you tap them briskly downward.

The patient will not be able to maintain extension and supination (and "drift" into pronation) with upper motor neuron disease.

Upper Motor Neuron (UMN) (see also Table 3.4)
- Descending motor tracts (pyramidal, extrapyramidal, cerebellar outflow etc.)
- Fiber tracts in transit from cerebrum, cerebellum, through brainstem into spinal cord until synapse with LMN

Lower Motor Neuron (LMN)
- Anterior horn cell
- Peripheral nerves

- Neuromuscular junction
- Muscle

Table 3.4 Neuroanatomic correlation of upper (UMN) and lower motor neuron (LMN) lesions

	UMN	LMN
Strength	Decreased	Markedly decreased
Tone	Increased	Decreased
Reflexes	Exaggerated	Decreased or absent
Atrophy	Mild	Significant
Fasciculation	Absent	Present
Babinski response	Present	Absent

Decerebrate Rigidity

In experimental animals **decerebrate rigidity** can be induced by transection of the brainstem at the superior border of the pons. The most prominent finding in **decerebrate rigidity** is marked spasticity of the body musculature. Decerebration produces no phenomenon akin to spinal shock, and the rigidity develops as soon as the brainstem is transected. Decerebrate rigidity is found to be spasticity due to diffuse facilitation of stretch reflexes. The facilitation is due to increased general excitability of the motor neuron pool and an increase in the rate of discharge in the gamma efferent neurons. The spasticity produced by decerebration is most marked in the extensor muscles (the muscles with which the body resists gravity).

Decorticate Rigidity

Decorticate rigidity is an abnormal posturing characterized by rigidity, flexion of the arms, clenched fists, and extended legs. The arms are bent inward toward the body with the wrists and fingers bent and held on the chest. Decorticate rigidity appears as a result of a lesion to the mesencephalon or above. Decorticate rigidity is seen on the hemiplegic side in humans after haemorrhages or thromboses in the internal capsule.

Coordination

Rapid Alternating Movements

Ask the patient to strike one hand on the thigh, raise the hand, turn it over, and then strike it back down as fast as possible.

Ask the patient to tap the distal thumb with the tip of the index finger as fast as possible.

Ask the patient to tap your hand with the ball of each foot as fast as possible.

Point-to-Point Movements

Ask the patient to touch your index finger and his nose alternately several times. Move your finger about as the patient performs this task.
Hold your finger still so that the patient can touch it with one arm and finger outstretched. Ask the patient to move his arm and return to your finger with his eyes closed.

Ask the patient to place one heel on the opposite knee and run it down the shin to the big toe. Repeat with the patient's eyes closed.

Romberg

Be prepared to catch the patient if he is unstable.

Ask the patient to stand with the feet together and eyes closed for 5–10 seconds without support.

The test is said to be positive if the patient becomes unstable (indicating a vestibular or proprioceptive problem).

Gait

Ask the patient to:
Walk across the room, turn and come back
Walk heel-to-toe in a straight line
Walk on their toes in a straight line
Walk on their heels in a straight line
Hop in place on each foot
Do a shallow knee bend
Rise from a sitting position

Reflexes

Deep Tendon Reflexes (Table 3.5)

The patient must be relaxed and positioned properly before starting.

Reflex response depends on the force of your stimulus. Use no more force than you need to provoke a definite response.

Reflexes can be reinforced by having the patient perform isometric contraction of other muscles (clenched teeth).

Reflexes should be graded on a 0 to 4 "plus" scale (Table 3.6)

Table 3.5 Examination of deep tendon reflexes

Reflex (root value)	Examination
Biceps (C5, C6)	The patient's arm should be partially flexed at the elbow with the palm down. Place your thumb or finger firmly on the biceps tendon. Strike your finger with the reflex hammer. You should feel the response even if you can't see it.
Triceps (C6, C7)	Support the upper arm and let the patient's forearm hang free. Strike the triceps tendon above the elbow with the broad side of the hammer. If the patient is sitting or lying down, flex the patient's arm at the elbow and hold it close to the chest. Watch for extension of arm.
Brachioradialis (C5, C6)	Have the patient rest the forearm on the abdomen or lap. Strike the radius about 1–2 inches above the wrist. Watch for flexion and supination of the forearm.
Abdominal (T8, T9, T10, T11, T12)	Use a blunt object such as a key or tongue blade. Stroke the abdomen lightly on each side in an inward and downward direction above (T8, T9, T10) and below the umbilicus (T10, T11, T12). Note the contraction of the abdominal muscles and deviation of the umbilicus towards the stimulus.
Knee (L2, L3, L4)	Have the patient sit or lie down with the knee flexed. Strike the patellar tendon just below the patella. Note contraction of the quadriceps and extension of the knee.
Ankle (S1, S2)	Dorsiflex the foot at the ankle. Strike the Achilles tendon. Watch and feel for plantar flexion at the ankle.

Table 3.6 Tendon reflex grading scale

0	Absent
1+ or +	Hypoactive
2+ or ++	Normal
3+ or +++	Hyperactive without clonus
4+ or ++++	Hyperactive with clonus

Clonus
If the reflexes seem hyperactive, test for ankle clonus:

Support the knee in a partly flexed position.

With the patient relaxed, quickly dorsiflex the foot.

Observe for rhythmic oscillations.

Plantar Response (Babinski)
Stroke the lateral aspect of the sole of each foot with the end of a reflex hammer or key.

Note movement of the toes, normally flexion (withdrawal).

Positive Babinski's is characterized by extension of the big toe with fanning of the other toes.

SENSORY EXAMINATION

Before proceeding for sensory examination explain each test to the patient. Compare symmetrical areas on the two sides of the body.

Compare distal and proximal areas of the extremities.

Map out any sensory loss and its boundaries in detail (Table 3.7).

Table 3.7 Important landmarks and root value

Root value	Area to be tested
C4	Shoulders
C6	Thumb and outer aspects of the forearms
C7	Middle finger
C8	Little finger and inner aspect of forearm
T1	Outer aspects of the forearms
T4	Nipple
T10	Umbilicus
L2	Front of both thighs
L3	Knee
S1	Sole
S5	Anus

Vibration

Use a low pitched tuning fork (128 Hz).
Test with a non-vibrating tuning fork first to ensure that the patient is responding to the correct stimulus.
Place the stem of the fork over the distal interphalangeal joint of the patient's index fingers and big toes.
Ask the patient to tell you if he feels the vibration.

If vibration sense is impaired proceed proximally:
Wrists
Elbows
Medial malleoli
Patellas
Anterior superior iliac spines
Spinous processes
Clavicles

Subjective Light Touch

Use your fingers to touch the skin lightly on both sides simultaneously.

Test several areas on both the upper and lower extremities.
Ask the patient to tell you if there is difference from side to side or other "strange" sensations.

Position Sense

Grasp the patient's big toe and hold it away from the other toes to avoid friction.
Show the patient "up" and "down."
With the patient's eyes closed ask the patient to identify the direction you move the toe.
If position sense is impaired move proximally to test the ankle joint.
Test the fingers in a similar fashion.
If indicated move proximally to the metacarpophalangeal joints, wrists, and elbows.

Dermatomal Testing

If vibration, position sense, and subjective light touch are normal in the fingers and toes you may assume the rest of this exam will be normal.

Pain

Use a suitable sharp object to test "sharp" or "dull" sensation.

Temperature

Often omitted if pain sensation is normal.
Use a tuning fork heated or cooled by water and ask the patient to identify "hot" or "cold."

Light Touch

Use a fine whisp of cotton or your fingers to touch the skin lightly.
Ask the patient to respond whenever a touch is felt.

Cortical Functions

Since these tests are dependent on touch and position sense, they cannot be performed when the tests above are clearly abnormal.

Graphesthesia
With the blunt end of a pen or pencil, draw a large number in the patient's palm.
Ask the patient to identify the number.

Stereognosis
Use as an alternative to graphesthesia.

Place a familiar object in the patient's hand (coin, paper clip, pencil, etc.). Ask the patient to tell you what it is.

Two Point Discrimination

Use in situations where more quantitative data are needed, such as following the progression of a cortical lesion.
Use an opened paper clip to touch the patient's finger pads in two places simultaneously.
Alternate irregularly with one point touch.
Ask the patient to identify "one" or "two."
Find the minimal distance at which the patient can discriminate.

PAEDIATRIC NEUROLOGICAL ASSESSMENT

Neurological assessment in infants and children consists of a thorough history beginning with conception and proceeding through pregnancy, birth, early neonatal course, subsequent hospitalizations, chronic medical conditions, acquisition of milestones in each category of normal development, medications, allergies, family history including consanguinity, social history, and review of systems (Table 3.8). One of the most important goals of a neurologic evaluation in children is to distinguish whether the neurological impairment represents a static condition (i.e., cerebral palsy) or a progressive process.

Table 3.8 Pediatric neurological history

Antenatal	Maternal parity Previous miscarriages or abortions Illnesses during the pregnancy Maternal nutrition and supplementation Medications taken during the pregnancy Alcohol/smoking Fetal movements Vaginal spotting or bleeding Premature rupture of the membranes
Perinatal	Spontaneous or induced labor Duration of labor Fetal monitoring during labor Fetal distress Estimated gestational age at time of delivery Apgar score Birth weight Head circumference
Neonatal complications	Jaundice Temperature Breathing Feeding (breast/bottle)

Neurodevelopment	Progressing or regressing Development of handedness Attainment of major milestones Academic performance in school
Immunizations	DPT, polio, BCG
Family history Social history	

Neurological Examination

The neurological examination must be tailored to the age and abilities of the infant or child. The routine newborn assessment should include an examination for size, macrocephaly or microcephaly, changes in skin colour, signs of birth trauma, malformations, evidence of respiratory distress, level of arousal, posture, tone, presence of spontaneous movements, symmetry of movements, and reflexes.

In younger patients, assessments often rely more on observation and play interactions than on direct questioning.

Apgar Score

The Apgar score is a practical method of systematically assessing newborn infants immediately after birth to help identify those requiring resuscitation and to predict survival in neonatal period (Table 3.9).

Table 3.9 APGAR score

Appearance	Blue: 0 Acrocyanotic: 1 Pink: 2
Pulse	Absent pulse: 0 Heart rate < 100 beats per minute: 1 Heart rate > 100 beats per minute: 2
Grimace (Reflex irritability)	No response: 0 Grimace: 1 Vigorous cough or sneeze: 2
Muscle tone	Limp: 0 Some flexion of extremities: 1 Active motion: 2
Respiratory effort	Respirations absent: 0 Slow or irregular breaths: 1 Good respiratory rate and cry: 2

Interpretation

Calculation (Maximum of 10 points)
One minute Apgar (term infants)
Apgar <5: Lower pH and higher $PaCO_2$
Apgar >7: Suggests stable oxygenation and perfusion
Five minute Apgar
Reflects infant's changing condition

Causes of Low Apgar Score

Asphyxia
Drugs (anaesthetics, sedatives, opiates)
Central nervous system diseases
Congenital muscular disease
Prematurity
Foetal sepsis

Head and Neck

Head circumference and fontanelle size can indicate a congenital disorder or head trauma.

Macrocephaly

Macrocephaly as an isolated anomaly, is often familial, with autosomal dominant inheritance; may be a manifestation of other anomalies, including hydrocephalus and skeletal disorders such as achondroplasia.

Microcephaly

Microcephaly can be familial, with autosomal dominant or recessive inheritance; may be associated with infections (viruses such as cytomegalovirus) and syndromes such as trisomy 13 and 18, Cornelia de Lange's, Rubinstein-Taybi, Prader-Willi, and foetal alcohol syndrome.

Caput Succedaneum

Caput succedaneum is commonly observed after prolonged labor. It is secondary to accumulation of blood or serum above the periosteum. Clinical examination shows poorly demarcated soft tissue swelling that crosses suture lines; accompanying pitting edema and overlying petechiae, ecchymoses and purpura. Usually it is treated conservatively and resolves within days.

Cephalhaematoma

Less common than caput succedaneum but may occur after prolonged labor and instrumentation. It is secondary to rupture of blood vessels that traverse skull to periosteum. Clinical examination shows a well-demarcated, often fluctuant swelling that does not cross suture lines; no overlying skin discolouration; possibly, skull fractures; sometimes, elevated ridge of organizing tissue.

Face

Facial nerve paralysis is caused by compression of the nerve against the sacral promontory or by trauma resulting from the use of forceps during delivery. Paralysis is usually apparent on the first or second day of life. The nasolabial fold on the paralyzed side is obliterated, and the corner of the mouth droops; with crying, the mouth is drawn to the normal side. Electrodiagnostic testing may be necessary if no improvement occurs within seven to 10 days; rarely, surgical intervention is needed.

The Motor Exam

Neurological exam may provide clues to motor impairment and these can be detected by keen observation. For example, any asymmetry of movement may suggest a focal CNS lesion and is important to note. Other clues to impaired motor function include:

Development of a hand preference before 12 months of age – indicative of motor impairment in the opposite side (hand preference typically does not develop until about 2 years of age).
Asymmetry of thumb-nail width, limb size or muscle mass – suggests hemiatrophy of the limb with motor impairment.
Persistent fisting of hands after 3 months of age – suggestive of hypertonia/cortical spinal tract dysfunction.
Presence of abnormal involuntary movements/limb posturing/ataxia – suggests injury to basal ganglia/cerebellum.
Marked hip abduction when lying supine – indicative of axial hypotonia.

Hypotonia (low muscle tone) is evidenced by a drooping of the child's head, trunk and limbs when the child is held in horizontal suspension, and by the child's shoulders slipping through the examiner's hands in vertical suspension. If hypertonia is present, scissoring of the lower extremities in vertical suspension may be noted.

Deep Tendon Reflexes

Elicitation of deep tendon reflexes requires practice and patience. In the presence of hypotonia, decreased or absent reflexes suggest a peripheral nervous system disease. Hyperreflexia in the presence of low or increased tone is indicative of CNS dysfunction. Other signs of upper motor neuron disease include ankle clonus (> 7–8 beats in infancy), a positive Babinski response (after 12 months of age), the Hoffman reflex, and the crossed adductor reflex.

Primitive Reflexes

Assessment of the primitive and protective postural reflexes (Table 3.10) is also a crucial part of the motor exam. Primitive reflexes normally develop in utero, while postural reflexes develop later in infancy. An asymmetry of response or abnormal persistence of primitive reflexes may be one of the earliest signs of abnormal motor development.

Table 3.10 Newborn reflexes

Reflex	Method
Foot	Stroke Inner Sole Toes curl around ("grasp") examiner's finger Stroke Outer Sole (Babinski) Toes spread, great toe dorsiflexion Asymmetry of the Babniski reflex between extrimities is suggestive of organic lesion
Doll's eyes	Give one forefinger to each hand - baby grasps both Pull baby to sitting with each forefinger Eyes open on coming to sitting (like a doll's) Head initially lags Baby uses shoulders to right head position
Walking reflex	Hold baby up with one hand across chest As feet touch ground, baby makes walking motion
Protective reflex	Soft cloth is placed over the baby's eyes and nose Baby arches head and turns head side to side Brings both hands to face to swipe cloth away
Rooting reflex	Touch newborn on either side of cheek Baby turns to find breast Sucking mechanism on finger is divided into 3 steps Front of tongue laps on finger Back of tongue massages middle of the finger Esophagus pulls on tip of finger
Tonic neck (fencing) reflex	If the Baby's head is rotated leftward The left arm (face side) stretches into extension The right arm flexes up above head Opposite reaction if head is rotated rightward
Moro reflex (startle reflex)	Hold supine infant by arms a few inches above bed Gently drop infant back to elicit startle Baby throws arms out in extension and baby grimaces Persistence of Moro's reflex beyond 12 weeks is pathognomonic of CNS pathology
Hand-to-Mouth (Babkin) reflex	Stroke newborns cheek or put finger in baby's palm Baby will bring his fist to mouth and suck a finger
Swimmer's (Gallant) response	Hold baby prone while supporting belly with hand Stroke along one side of baby's spine Baby flexes whole body toward the stroked side
Crawling reflex	Newborn placed on abdomen Baby flexes legs under him and starts to crawl

NORMAL CHILD DEVELOPMENT

Normal development is assessed in the categories gross motor, fine motor, speech and language, and social/adaptive (Table 3.11).

Developmental Milestones

Cognitive Milestones

Month 3–5: Attends to and reaches for objects
Month 4–8: Pulls string to secure a ring
Month 8–15: Imitates patting doll
Month 14–20: Finds hidden object
Month 18–28: Completes simple puzzles

Language Milestones

Month 1.5–3: Squeals
Month 3.5–8: Turns to locate a voice
Month 9–13: Says Mama or Dada
Month 14–24: Combines two different words
Month 21–36: Uses plurals

Social and Emotional Milestones

Month 1.5–4: Smiles at others
Month 4–9: Seeks primary caregiver
Month 8–15: Stranger anxiety
Month 10–15: Displays 2 or more recognizable emotions
Month 11–20: Exploratory play by self
Month 21–36: Cooperative play in small groups

Gross Motor Milestones

Month 2–4.5: Rolls over
Month 5–8: Sits without support
Month 10–14: Stands alone
Month 14–20: Walks up steps
Month 21–28: Pedals tricycle
Month 30–44: Balances on one foot
By age 6: Rhythmic skipping
By age 8.5: Alternates foot-hop in place
By age 10: Holds tandem stance for 10 sec (eyes closed)

Fine Motor Milestones

Month 2.5–4: Grasps rattle
Month 4.5–7: Transfers cube hand to hand
Month 8–12: Has neat pincer grasp

Table 3.11 Normal development of language, perception, and motor skills

Average age	Language achievements	Perceptual achievements	Motor achievements
Gestational		Can hear sounds	Can swallow, suck thumb, move limbs
Infancy			
Birth–3 months	Understands phenomes; responds to sound; can distinguish speech sounds; Chomsky's language acquisition	Sound, touch, smell, taste present in newborns; sees patterns and shape, contrast	Balances head; lifts by arms
2 months	Cooing; turn taking with caregivers	All perceptions more acute	Rolls side to side; reaches for object
28–40 weeks	Knows friendly from unfriendly voices Native language discrimination	Seeing more focused; sees all colours and patterns; vision and hearing coordinated Responds to motion; size constant; sees depth	Sits, pokes; crawls, plays pat-a-cake
6 months	Babbling; self-expressions for needs		
40 weeks–year	Words connected to meaning; first words	Can see features. Child sees at 20/100 at 6 months	Stands, walks, builds

contd.

1½–2½ years	First sentence—two words; 50 word vocabulary	Tracks objects	Scribbles
Early childhood			
6–11 years	Metalinguistics understood; child improves inflection, pronouncement		Boys better gross motor skills; girls overall more developed
Adolescence			
11 years and older			Gains in gross motor performance; girls' gross motor performance tapers at age 14; boys spurt through teens

Month 15–20: Builds tower of four cubes
Month 18–24: Imitates vertical line
Month 28–36: Copies circle
By age 5 years: Draws a square
By age 5.5 years: Tripod pencil grasp
By age 7 years: Draws diagonal line
By age 9: Draws cross with same dimensions
By age 12: Draws three dimensional cube

CHAPTER 4

Lumbar Puncture

INDICATIONS FOR LUMBAR PUNCTURE

Central nervous system (CNS) infections (meningitis, and encephalitis)
Subarachnoid haemorrhage
Confusional states
Acute stroke
Status epilepticus
Meningeal malignancies
Demyelinating diseases
CNS vasculitis
Lumbar puncture has limited therapeutic usefulness, for example, intrathecal therapy in meningeal malignancies and fungal meningitis.

CONTRAINDICATIONS FOR LUMBAR PUNCTURE

Presence of infection in the skin overlying the spine
Suspected intracranial mass lesions
Suspected brain herniation syndromes (for example, uncal, cerebellar, or cingulate herniation)
Papilloedema
Thrombocytopenia and other bleeding diatheses
The availability of CT has simplified the management of patients with papilloedema. If CT reveals no evidence of a mass lesion, then lumbar puncture is usually needed in the presence of papilloedema to establish the diagnosis of pseudotumour cerebri and to exclude meningeal inflammation or malignancy.

COMPLICATIONS OF LUMBAR PUNCTURE

Headache
- Most common complication
- Results from low cerebrospinal fluid pressures due to persistent fluid leakage through the dural hole
- Pain is present in the upright position and is promptly relieved with a supine position
- The headache is aggravated by cough or strain.
- Occasionally it is associated with nausea, vomiting, or tinnitus

Worsening of brain herniation and spinal cord compression
Subarachnoid bleeding
Diplopia
Backache

CEREBROSPINAL FLUID PRESSURE

The cerebrospinal fluid pressure should be measured routinely.

Causes of Low Pressures
- Dehydration
- Spinal subarachnoid block
- Following previous lumbar puncture
- Other cerebrospinal fluid leaks (rhinorrhoea, otorrhoea)
- Faulty needle placement

Causes of Increased Pressure
- Brain oedema
- Infections
- Acute stroke
- Cerebral venous occlusions
- Congestive heart failure
- Pulmonary insufficiency
- Benign intracranial hypertension (pseudotumour cerebri)

CEREBROSPINAL FLUID FINDINGS

Normal cerebrospinal fluid contains no more than five lymphocytes or mononuclear cells/mm^3. A higher white cell count is pathognomonic of disease in the central nervous system or meninges. The changes characteristic of the various meningitides are listed in Table 4.1.

BLOOD IN THE SUBARACHNOID SPACE

Needle trauma
Acute subarachnoid haemorrhage

Table 4.1 Cerebrospinal fluid findings in meningitis

Meningitis	Pressure (mm H_2O)	Leucocytes/mL	Protein (g/L)	Glucose (mmol/L)
Acute bacterial	Usually elevated	Several hundred to more than 60 000, usually a few thousand; occasionally less than 100 (especially meningococcal or early in disease); polymorphonuclears predominate	Usually 1 to 5, occasionally more than 100	2 to 2.2 in most cases, in absence of hyperglycaemia
Partially treated bacterial	Usually elevated; may be low with dynamic block in advanced stages	Usually 25 to 100; rarely more than 500; lymphocytes predominate except in early stages when polymorphonuclears may account for 80 percent of cells	Nearly always elevated, usually 1 to 2; may be much higher in dynamic block	Usually reduced, to 2.2 or less in 3/4 cases
Pseudomonal	Usually elevated	0 to 800; average 50; lymphocytes predominate	Usually 0.2 to 5, average 1	Reduced in most cases, average 1.7 (in absence of hyperglycaemia)
Viral	Normal to moderately elevated	5 to a few hundred; but may be more than 1000, particularly with lymphocytic choriomeningitis; lymphocytes predominate but may be more than 80 percent polymorphonuclears in first few days	Frequently normal or slightly elevated; less than 1, may show greater elevation in severe cases	Normal (reduced in 1/4 cases of mumps and herpes simplex)

cont.

Syphilitic (acute)	Usually elevated	Average 500; usually lymphocytes; rarely polymorphonuclear	Average, 1	Normal (rarely reduced)
Cysticercosis	Often increased; low with dynamic block	Increased mononuclears and polymorphonuclears with 2 to 7 percent eosinophilia in about half the cases	Usually 0.5 to 2	Reduced in 1/5 cases
Sarcoid	Normal to considerably elevated	0 to less than 100 mononuclear cells	Slight to moderate elevation	Reduced in 1/2 cases
Tuberculous	Normal or elevated	0 to several hundred mononuclears plus malignant cells	Elevated often to high levels	Normal or greatly reduced (low in 3/4 carcinomatous meningitis cases)

CHAPTER 5

Radiological Investigations

Radiology and other forms of medical imaging are fundamental to the diagnosis of central nervous system diseases. However, the final element in diagnosis and characterization of disease process needs correlation of clinical findings. These investigations are discussed separately for intracranial and spinal disorders.

INTRACRANIAL DISORDERS

Plain Films

The ready availability of CT in most neuroradiological units has led to a re-evaluation of the role of conventional skull radiographs, though their importance in craniofacial trauma and in the assessment of pituitary and skull-based lesions remains undiminished (Box 5.1).

Box 5.1 Basic radiographic projections

- Lateral view
- Posteroanterior view
- Towne's view
- Submentovertical (basal) view

Computed Tomography (CT)

In CT, film is replaced by sensitive detectors which move synchronously with a collimated x-ray beam directed at the edge of a narrow slice of tissue. Multiple precise measurements of the amount of transmitted radiation are processed by computer to produce information about very small volumes of tissue (voxels) within the slice. The image can be viewed on a monitor and printed also.

Advantages

Discrimination is possible between white and grey matter, cerebral ventricles, and the cerebrospinal fluid pathways.

In the orbit the optic nerve, extraocular muscles, and other retrobulbar structures are better identifiable due to the surrounding low attenuation fat.

Calcification is well demonstrated
Detection of intracranial haematomas (extradural, subdural etc.)
Detection of cerebral infarction
Intravenous injections of an iodine-containing contrast medium increases the sensitivity of CT and helps to detect brain tumours, pituitary tumours.
Easy to use and non-invasive nature
Can be performed as an outpatient procedure

Disadvantages

Radiation exposure
Poor delineation of posterior fossa and spinal cord

Magnetic Resonance Imaging (MRI)

MRI employs radiofrequency radiation in the presence of a magnetic field to produce images. Intravenous contrast agents containing gadolinium increase the sensitivity of MRI

Advantages

More sensitive imaging modality than CT scan to demonstrate cerebral disease processes (Box 5.2)
Investigation of choice to image posterior fossa and spine
No radiation

Box 5.2 MRI is highly sensitive in detecting

Infective and inflammatory changes in cerebral tissue
Encephalitic
Demyelinating infarcts in multiple sclerosis
Cerebral tumours
Posterior fossa
Acoustic neuromas

Disadvantages

Uncertainty in the detection of soft-tissue calcification

Box 5.3 MRI contraindications

Cardiac pacemakers
Small metallic foreign bodies, including some surgical clips

Magnetic Resonance Angiography (MRA)

MRA provides a non-invasive and radiation-free technique for the investigation of intra- and extracerebral vascular structures without the use of contrast agents based on the movement of protons in flowing blood in blood vessels. However, morphological detail remains inferior to conventional angiography and is, as yet, inadequate for the reliable investigation of any aneurysmal disease or for the planning of endovascular therapy.

Cerebral Angiography

Cerebral angiography is used to visualize the intracranial circulation. This can be achieved either by percutaneous puncture or selective catheterization of the appropriate carotid or vertebral artery and injection of iodinated contrast. It can be used to diagnose aneurysm, vascular malformations and to know the blood supply of certain tumours (i.e., meningioma).

Radio-isotpe Nuclear Imaging (RNI)

Radiopharmaceuticals (99 Tc-Technetium pertechnetate) administered intravenously is the most commonly used intravenous agent for static brain studies. Detectors in a rectilinear scanner or gamma-camera register activity for normal soft tissue, the major venous sinuses, and some pathological processes within the brain. Only those abnormalities which are vascular or destroy the blood-brain barrier are demonstrated

Advantages

Safe
Little radiation dose
Can be carried out as an outpatient procedure

Disadvantages

Low sensitivity
High false negative rate
Lack of tissue characterization

Single Proton Emission Computed Tomography (SPECT)

SPECT uses computer reformation techniques similar to those employed in CT. In this technique cerebral agent labelled with single-photon short-lived

gamma-emitters (iodo-isopropyl amphetamine [IMP] and hexa-methyl-propoline-amine [HM-PAO]), which cross the blood-brain barrier, provides a means of imaging regional cerebral metabolism.

Uses

HM-PAO uptake has been used to identify increased neuronal activity responsible for focal epilepsy
To differentiate the variety of atrophic dementing disorders
As a confirmatory technique in the assessment of brain death

Ventriculography

Demonstration of the cerebral ventricular system by the introduction of air, iodized oil, or water-soluble iodinated contrast material through a burr hole. This has now been completely replaced by CT and MRI.

Ultrasound

Ultrasonic vibrations are produced by passing an electric current through a suitable crystal (Pizzo-electric crystal). The sound-waves pass directly through homogeneous matter but are reflected back by certain interfaces and detected by the same crystal. Ultrasound scan can demonstrate midline intracranial structures, ventricular size, ventricular walls and subdural collections in infants as their fontanelles are open. Ultrasound does not employ ionizing radiation and has no hazard but does require considerable operator skill.

SPINAL DISORDERS

Plain Films

Conventional radiography may reveal congenital (i.e., atlanto-axial dislocation) and acquired bone abnormalities (fracture dislocation), together with indirect evidence of intervertebral disc disease (reduced disc space scoliosis).

Isotope Bone Scanning

In isotope scanning most commonly used substance is technetium labelled phosphate complex (methylene diphosphonate). It is a primary screening method in the study of metastatic bone disease.

Myelography

In myelography a radiographic contrast material into the spinal subarachnoid space is introduced. Contrast can be introduced by the lumbar or cisternal route. Non-ionic, low osmolarity agents which, because of their water solubility and low density, provide a clear outline of the nerve roots and intrathecal contents. With the availability of MRI it is not commonly used.

Computed Tomography

CT is a non-invasive procedure and provides an axial projection of spinal topographical anatomy. It provides a more precise identification of the articular configuration of the apophysial joints and their relation to the spinal canal and intervertebral foramina. CT is used in the evaluation of bony anomalies (spine fractures and cranio-vertebral junction anomalies).

Magnetic Resonance Imaging

MRI is the investigation of choice for the assessment of spine and spinal cord disorders.

Advantages

Non-invasive
No radiation dose
Allows sectional imaging in multiple planes
Intramedullary lesions are directly visualized by MRI and better delineation of the extent
Can differentiate between syrinx, tumour cyst, and solid tumour
Define the relationship of tumours (intra or extramedullary) to the spinal cord and the root
Choice of investigation in degenerative spinal disease

Spinal Angiography

Spinal angiography is valuable in the diagnosis of spinal arteriovenous malformations and of arteriovenous dural fistulae. It has limited role in the diagnosis of spinal tumours

CHAPTER 6

Electrodiagnostic Investigations

ELECTROENCEPHALOGRAPHY

The electroencephalogram (EEG) assesses functional aspects of brain and its most effective use, other than in screening, is where there are no, or inadequately demonstrable, structural abnormalities.

The EEG relates to the brain roughly as the ECG relates to the heart and its use is similar. The recordings are measured in microvolts (mV), and require sophisticated equipment and recording techniques to avoid artefacts and extraneous influences. Interpretation of EEG requires an intimate knowledge of the technology as well as clinical presentations.

For practical purposes the EEG can be assumed to be generated only by the neurones of the cerebral cortex. Non-neuronal elements, tumours, blood clots, infarcts, cysts, and abscesses, are intrinsically silent.

Waves

Alpha Rhythm

Alpha rhythm refers to a [illegible] sinusoidal sequence of waves at between 8 and 13 cycles/s seen in occipital areas when [illegible] in the resting state. This may be slowed in a variety of [illegible] conditions and locally.

Beta Rhythm

Beta rhythms are fast frequencies, greater than 13 cycles/s, seen [illegible] in anterior and central regions. Beta activity is enhanced by sedative drugs and can be localized for superficial and chronic lesions.

Delta Waves

Delta waves, or rhythms, if they are repetitive, are slow waves, less than 4 cycles/s. They are always abnormal in the alert adult and commonly associated with pathology, but normal in sleep, infants, children, and young adolescents.

Theta Waves

Theta waves, or intermediate slow frequencies, 4 to 8 cycles/s, are very variable, occur commonly in the temporal regions, and decline with maturity. They are often of pathological significance.

Advantages

Relatively inexpensive
Simple
Non-invasive
Can be carried out at the bedside

Limitations

Needs experience and clinical acumen for interpretation of the record
The majority of EEG abnormalities are non-specific and have many differential possibilities.

Uses

Computed tomography (CT) scanning and magnetic resonance imaging (MRI) have taken the place of EEG as a noninvasive means of screening for focal structural abnormalities of the brain, such as tumours, infarcts, or haematoma.

Epilepsy

The EEG is most useful in evaluating patients with suspected epilepsy (presence of electrographic seizure activity, i.e., of abnormal, repetitive, rhythmic activity). The absence of electrocerebral abnormalities does not exclude a seizure disorder.

Coma

The EEG tends to become slower as consciousness is depressed, regardless of the underlying cause. The EEG is useful [illegible] patients with locked-in syndrome and helps in distinguishing this disorder from the comatose state.

EVOKED POTENTIALS

Sensory Evoked Potentials

The noninvasive recording of spinal or cerebral potentials elicited by stimulation of specific afferent pathways is an important means of monitoring the functional integrity of these pathways but does not indicate

the pathologic basis of lesions involving them. These potentials are recorded from scalp or neck electrodes at a considerable distance from their generators in the spinal cord, brainstem or cerebral hemispheres, the voltages generated are extremely small, generally of the order of 0.25 and 25 mV. As evoked potentials provide an objective method of testing sensory function, it can be used to investigate patients unable or unwilling to co-operate.

Visual Evoked Potentials (VEPs)

Visual evoked potentials are elicited by monocular stimulation with a reversing checkerboard pattern and are recorded from the occipital region in the midline and on either side of the scalp. VEPs are most useful in detecting dysfunction of the visual pathways anterior to the optic chiasma (e.g., acute severe optic neuritis, optic nerve disease, such as ischaemia or compression by a tumour).

Somatosensory Evoked Potentials (SEPs)

The somatosensory evoked potential (SEP) is usually evoked by an electrical stimulus applied transcutaneously to a peripheral nerve, either in the upper limb (the median or ulnar nerve at the wrist or elbow are commonly used), the lower limb (posterior tibial nerve at the ankle or lateral popliteal at the knee), or, less commonly, the face (trigeminal nerve). The configuration, polarity, and latency of the responses depend on the nerve that is stimulated and on the recording arrangements. SEPs are used to evaluate proximal (otherwise inaccessible) portions of the peripheral nervous system and the integrity of the central somatosensory pathways. They have been used particularly to investigate patients with suspected multiple sclerosis (MS), investigation and monitoring of traumatic injuries to the peripheral nerves, spinal roots, or the cord, intraoperative monitoring etc. In patients in coma, prolongation of the central conduction time may provide a useful warning of incipient ischaemic damage.

Auditory Evoked Potential

The auditory evoked potential consists of seven subcortical components generated by the arrival of the afferent sensory volley at or near the following points on the auditory pathway: the auditory nerve (wave I), the cochlear nucleus (wave II), the superior olivary complex (wave III), the lateral lemniscus (wave IV), the inferior colliculus (wave V), the medial geniculate (wave VI), and the geniculocortical fibres (wave VII).

Clinical Uses

Testing hearing in infants and older children or adults who are unable or unwilling to cooperate in conventional audiometry

Investigation of patients with suspected demyelinating disease or other lesions of the central auditory pathways.

Early detection of acoustic neuromas.

Motor Evoked Potentials

The electrical potentials recorded from muscle or the spinal cord following stimulation of the motor cortex or central motor pathways are referred to as motor evoked potentials. This may provide information of prognostic relevance (for example, in suggesting the likelihood of recovery of motor function after stroke) and may be useful as a means of monitoring intraoperatively the functional integrity of central motor tracts.

ELECTROMYOGRAPHY (EMG)

The pattern of electrical activity in muscle (i.e., the electromyogram [EMG]), both at rest and during activity, may be recorded from a needle electrode inserted into the muscle. It aims to detect, and distinguish between, disorders of anterior horn cell, root, plexus, peripheral nerve, neuromuscular junction, and muscle, to determine their extent and severity, and to relate neurophysiological abnormality to the clinical context in which it is found. EMG enables disorders of the motor units to be detected and characterized as either neurogenic or myopathic. In neurogenic disorders, the pattern of affected muscles may localize the lesion to the anterior horn cells or to a specific site as the axons traverse a nerve root, limb plexus, and peripheral nerve to their terminal arborizations. Electromyography can neither prove nor disprove the existence of an upper motor neuron lesion.

NERVE CONDUCTION STUDIES

Nerve conduction studies complement the EMG examination, enabling the presence and extent of peripheral nerve pathology to be determined. They are particularly helpful in determining whether sensory symptoms are arising from pathology proximal or distal to the dorsal root ganglia (in the former instance, peripheral sensory conduction studies will be normal) and whether neuromuscular dysfunction relates to peripheral nerve disease. In patients with a mononeuropathy, they are invaluable as a means of localizing a focal lesion, determining the extent and severity of the underlying pathology, providing a guide to prognosis, and detecting subclinical involvement of other peripheral nerves.

Section II

DISORDERS OF BRAIN

CHAPTER 7

Stroke

STROKE/CEREBROVASCULAR DISEASE

Cerebrovascular disease includes disorders of the vascular system which cause ischaemia, infarction or haemorrhage in the brain.

Transient ischaemic attack is an acute loss of focal cerebral or monocular function with symptoms lasting less than 24 h and usually after adequate investigation, is presumed to be due to embolic or thrombotic vascular disease.

A stroke (or cerebrovascular accident) is a rapidly developing episode of focal, and at times global (applied to patients in deep coma and to those with subarachnoid haemorrhage), loss of cerebral function with symptoms lasting more than 24 h or leading to death, with no apparent cause other than that of vascular origin; the main pathological types of stroke are cerebral infarction, primary intracerebral haemorrhage, and subarachnoid haemorrhage.

Epidemiology

Cerebral infarction—80%
Primary intracerebral haemorrhage—10%
Subarachnoid haemorrhage—10%

Risk Factors

Hypertension
Heart disease of any kind
Atrial fibrillation
Transient ischaemic attack
Peripheral vascular disease

Diabetes mellitus
Smoking
High blood cholesterol
High blood fibrinogen
Factor VII coagulant activity
Heavy alcohol consumption
Contraceptive pills

Transient ischaemic attacks

Transient ischaemic attacks are characterized by focal ischaemic cerebral neurologic deficits that last for less than 24 hours (usually less than 1–2 hours).

Aetiology

An important cause of transient cerebral ischaemia is embolization.

Source of Emboli
Rheumatic heart disease
Mitral valve disease
Cardiac arrhythmia
Infective endocarditis
Atrial myxoma
Mural thrombi complicating myocardial infarction

Clinical Findings

The symptoms of transient ischaemic attacks vary markedly [illegible]
Onset of symptoms is usually abrupt and without warning [illegible] usually occurs rapidly, often within a few minutes.

Carotid Territory Ischaemia

Symptoms
Weakness and heaviness of the contralateral arm, leg or face, or [illegible] any combination

Numbness or paraesthesias with or without motor deficit

Slowness of movement, dysphasia, or monocular visual loss in [illegible] contralateral to affected limbs.

Neurological Examination
Flaccid weakness with pyramidal distribution
Sensory changes

Hyperreflexia
Extensor plantar response on the affected side, dysphasia
These findings may be present alone or in combination.
Subsequently, examination reveals no neurologic abnormality.

Vertebrobasilar Territory Ischaemia

Vertigo, ataxia, diplopia, dysarthria, dimness or blurring of vision, perioral numbness and paraesthesias, and weakness or sensory complaints on one, both, or alternating sides of the body
These symptoms may occur singly or in any combination
Drop attacks due to bilateral leg weakness without headache or loss of consciousness, may occur, sometimes in relation to head movements.

Neurological Examination
Signs of weakness, speech impairment, or gait disturbance either alone or in combination
Hyperreflexia
Extensor plantar response on the affected side, dysphasia
Subsequently, examination reveals no neurologic abnormality.

Differential Diagnosis

Migraine
Focal epileptic seizures

Rare causes
Brain tumour
Subdural haematoma
Arteriovenous malformation
Giant aneurysm
Multiple sclerosis
Labyrinthine disorders

Investigations

Clinical and laboratory evaluation must include assessment for hypertension, heart disease, haematologic disorders, diabetes mellitus, hyperlipidaemia, and peripheral vascular disease.
Complete blood count
Fasting blood glucose
Serum cholesterol
ECG
Chest X-ray
Echocardiography

Imaging

CT scan of the head to exclude the presence of intracerebral haemorrhage or a cerebral tumour

Ultrasonography

To study the cerebral circulation

Carotid duplex ultrasonography

To detect stenosis of the internal carotid artery

Arteriography

Most reliable

Will demonstrate the status of the cerebrovascular system

MR angiography may reveal stenotic lesions of large vessels more sensitive than conventional arteriography.

Treatment

Medical Treatment

Medical treatment is aimed at preventing further attacks and strokes

Stop smoking

Treat cardiac sources of embolization

Control of hypertension and diabetes

Treat hyperlipidemia

Anticoagulants.

Anticoagulants (Intravenous heparin followed by oral anticoagulants) are indicated for the treatment of embolism from the heart

Surgical Treatment

Removal of any tightly stenosing atherothrombotic lesion at the origin of the symptomatic internal carotid artery (High-grade stenosis of carotid artery (70–99% in luminal diameter.)

STROKE

Stroke is one of the leading causes of morbidity and mortality. Strokes can be subdivided pathologically into infarcts (thrombotic or embolic) and haemorrhages, and clinical criteria for distinguishing between these possibilities have been emphasized. However it is often difficult to determine on clinical grounds the pathological basis for stroke.

Lacunar Infarction

Lacunar infarcts are small lesions (usually 3–15 mm in diameter) involved in the distribution of single penetrating arteries arising from the basilar or cerebellum, anterior limb of the internal capsule, and less commonly the

deep cerebral white matter. Lacunar infarcts are associated with poorly controlled hypertension or diabetes and have been found in several clinical syndromes, including contralateral pure motor or pure sensory deficit, ipsilateral ataxia with crural paresis, and dysarthria with clumsiness of the hand. The neurologic deficit may progress over 24–36 hours before stabilizing.

Lacunar infarcts are sometimes visible on CT scans as small, punched-out, hypodense areas, but in other patients no abnormality is seen (Figure 7.1). In some instances, patients with a clinical syndrome suggestive of lacunar infarction are found on CT scanning to have a severe hemispheric infarct.

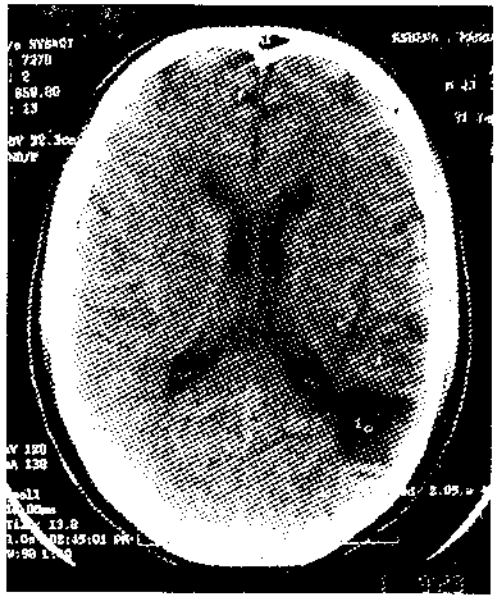

Fig. 7.1 Small lacunar infarction involving left parietal cortex

The prognosis for recovery from the deficit produced by a lacunar infarct is usually good, with partial or complete resolution occurring over the following 4–6 weeks in many instances.

Cerebral Infarction

Thrombotic or embolic occlusion of a major vessel leads to cerebral infarction. Causes include the disorders predisposing to transient ischaemic attacks (see above) and atherosclerosis of cerebral arteries. The resulting deficit depends upon the particular vessel involved and the extent of any collateral circulation.

Clinical Features

Onset is usually abrupt, and there may then be very little progression except that due to brain swelling. Clinical evaluation should always include examination of the heart and peripheral vessels.

Obstruction of Carotid Circulation
Occlusion of the Ophthalmic Artery
Most of the time asymptomatic because of rich collaterals
Transient embolic obstruction leads to amaurosis fugax (i.e., sudden and brief loss of vision in one eye).

Occlusion of the Anterior Cerebral Artery
Weakness and cortical sensory loss in the contralateral leg
Mild weakness of the arm (especially proximally)
Contralateral grasp reflex
Paratonic rigidity
Abulia (lack of initiative) or frank confusion
Urinary incontinence is not uncommon

Middle Cerebral Artery Occlusion
Contralateral hemiplegia
Hemisensory loss
Homonymous hemianopia (i.e., bilaterally symmetric loss of vision in half of the visual fields)
Deviation of eyes to the side of the lesion
If the dominant hemisphere is involved, global aphasia is also present.

Obstruction of Vertebrobasilar Circulation
Occlusion of the posterior cerebral artery
Contralateral hemisensory disturbances
Development of spontaneous pain and hyperpathia
Homonymous hemianopia with macular-sparing
Mild, usually temporary, hemiparesis. Depending on the site of the lesion and the collateral circulation, the severity of occlusion
Vertebral artery occlusion distally, below the origin of the anterior spinal and posterior inferior cerebellar arteries, may be clinically silent because the circulation is maintained by the other vertebral artery.

Basilar Artery Occlusion
Contralateral hemiplegia and sensory deficit
Ipsilateral cranial nerve palsy at the level of the lesion.

Occlusion of Both Vertebral Arteries
Coma with pinpoint pupils, flaccid quadriplegia and sensory loss, and variable cranial nerve abnormalities.

Partial basilar artery occlusion, there may be diplopia, visual loss, vertigo, dysarthria, ataxia, weakness or sensory disturbances in some or all of the limbs, and discrete cranial nerve palsies. In patients with hemiplegia of pontine origin, the eyes are often deviated to the paralyzed side, whereas in patients with a hemispheric lesion, the eyes commonly deviate from the hemiplegic side.

Laboratory and Other Studies

Complete blood count
Erythrocyte sedimentation rate
Blood glucose
Serologic tests for syphilis
Antiphospholipid antibodies (lupus anticoagulants and anticardiolipin antibodies)
Blood cultures should be performed if endocarditis is suspected
Echocardiography if heart disease is suspected

Imaging

X-ray Chest
Cardiomegaly or valvular calcification

CT Scan
CT scan of the head (without contrast) to exclude cerebral haemorrhage (Figure 7.2 A–C)

MRI and MR angiography
MRI in the acute stages may not be possible however diffusion-weighted MRI is more sensitive than standard MRI in detecting cerebral ischaemia.

Carotid Doppler Studies
Conventional Angiography

Treatment

Anticoagulants
If the neurologic deficit progresses over the following minutes or hours, heparinization may be of value in limiting or arresting further deterioration.

Clinical Features

Onset is usually abrupt, and there may then be very little progression except that due to brain swelling. Clinical evaluation should always include examination of the heart and peripheral vessels.

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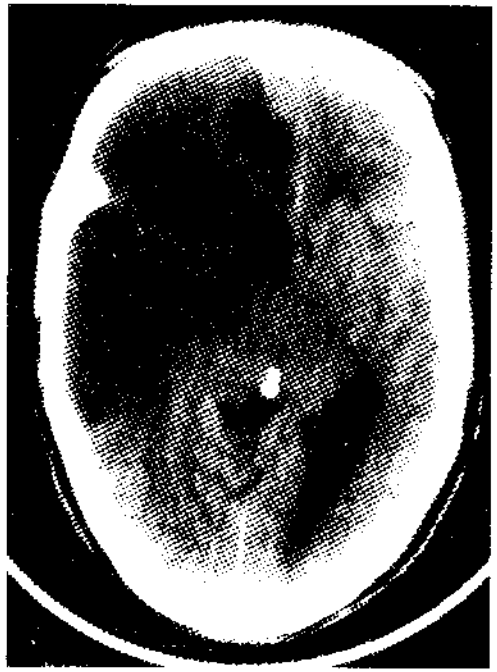

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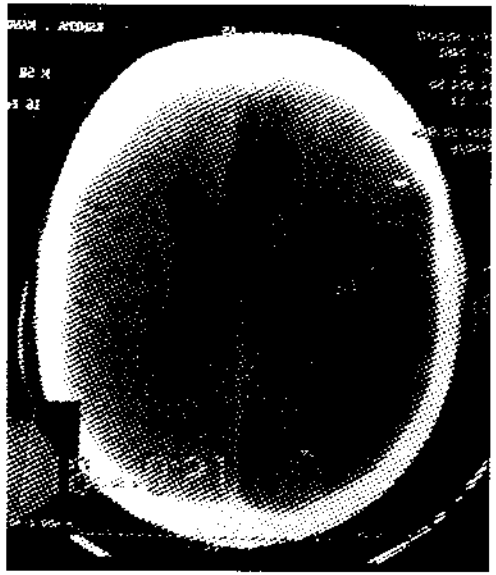

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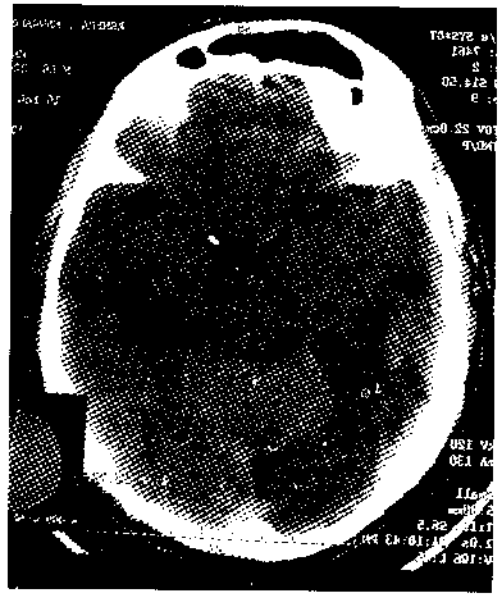


Fig. 7.2 CT scan images with patients of cerebral infarction

A. Ischaemic infarction involving right middle and anterior cerebral artery territory

B. Ischaemic infarction suggestive of left internal carotid artery occlusion

C. Ischaemic infarction involving the left posterior cerebral artery territory

Intravenous Thrombolytic Therapy

Intravenous thrombolytic therapy with recombinant tissue plasminogen activator is effective in reducing the neurologic deficit in selected patients without CT evidence of intracranial haemorrhage when administered within 3 hours after onset of ischaemic stroke

Control of Blood Pressure and Diabetes
General Supportive Measures
Management of Intracranial Pressure

Corticosteroids

Prednisone (up to 100 mg/d) or dexamethasone (16 mg/d)

Mannitol
Physical Therapy
Physical therapy has an important role in the management of patients with impaired motor function.

Passive movements at an early stage will help prevent contractures.

As cooperation increases and some recovery begins, active movements will improve strength and coordination.

Early mobilization and active rehabilitation are important.

Occupational therapy may improve morale and motor skills, while speech therapy may be beneficial in patients with expressive dysphasia or dysarthria.

When there is a severe and persisting motor deficit, a device such as a leg brace, toe spring, frame, or cane may help the patient move about, and the provision of other aids to daily living may improve the quality of life.

Intracerebral Haemorrhage

Intracerebral haemorrhages usually occur suddenly and without warning, often during activity. Bleeding can occur primarily into the subarachnoid space (i.e., aneurysm) or into the parenchyma (hypertension).

Causes

Aneurysm
Angioma
Hypertension
Haematologic and bleeding disorders (e.g., leukaemia, thrombocytopenia, haemophilia, or disseminated intravascular coagulation)
Anticoagulant therapy
Liver disease
Primary or secondary brain tumours

Clinical Features

Cerebral Haemorrhage (Figure 7.3 A–C)
Loss of consciousness:
With haemorrhage into the cerebral hemisphere, consciousness is initially lost or impaired in about one-half of patients.
Vomiting and headache:
Vomiting occurs very frequently at the onset of bleeding, and it is associated with headache.

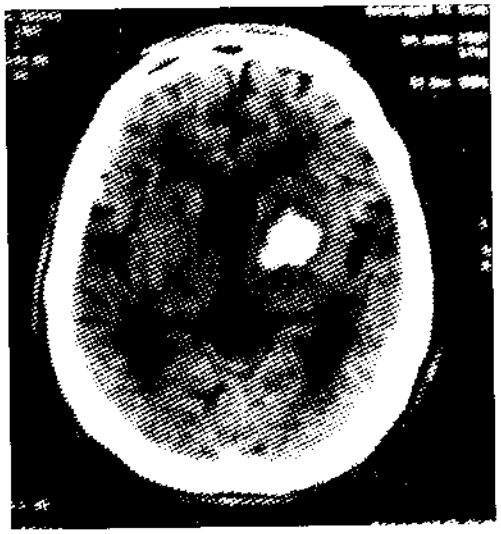
Focal symptoms and signs:
Focal symptoms and signs depend on the site of the haemorrhage.
There is generally a rapidly evolving neurologic deficit with hemiplegia or hemiparesis. A hemisensory disturbance is also present with more deeply placed lesions.
With lesions of the putamen, loss of conjugate lateral gaze may be conspicious.
With thalamic haemorrhage, there may be a loss of upward gaze, downward or skew deviation of the eyes, lateral gaze palasies, and pupillary inequalities.

Cerebellar Haemorrhage (Figure 7.4, 7.5 A and B)
Sudden onset of nausea and vomiting
Disequilibrium
Headache
Loss of consciousness
May terminate fatally within 48 hours

Differential Diagnosis

Chronic subdural haematoma
Drug overdose, in an unconscious patient with few or no focal signs, and without a good history of a sudden onset
Encephalitis, cerebral abscess, sudden deterioration in a patient with a cerebral tumour, multiple sclerosis, peripheral nerve lesion, hypoglycaemia, and somatization can all usually be excluded after an adequate history, clinical examination, and straightforward investigations.

A



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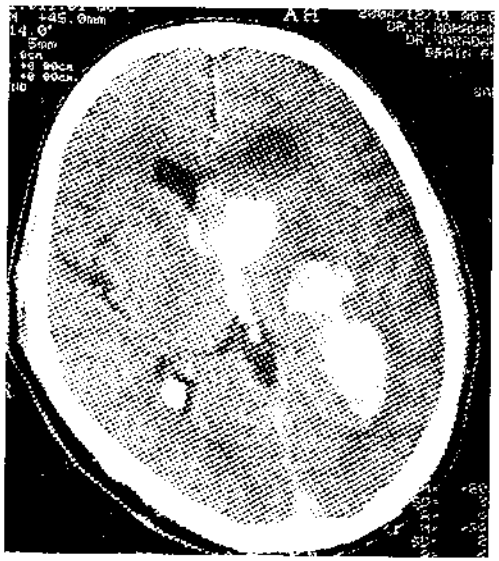

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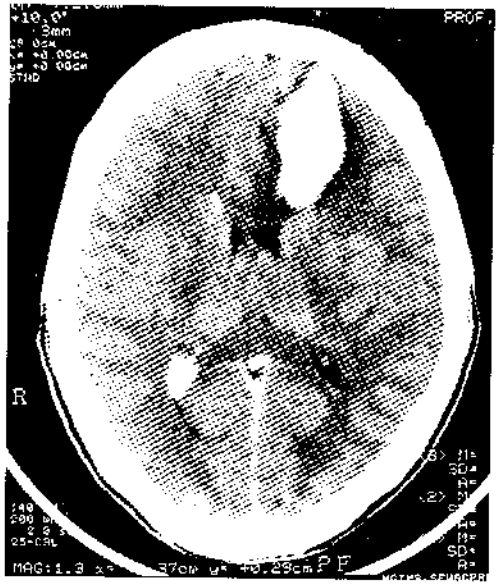

Fig. 7.3 CT scan images of cerebral haemorrhage

A. Small left basal ganglionic haematoma

B. Massive left thalamic and basal ganglionic haematoma with intraventricular extension

C. Left frontal lobe haematoma

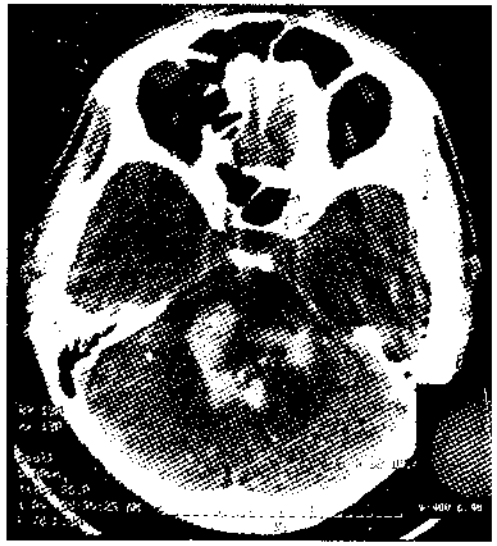

Fig. 7.4 CT scan showing haemorrhage involving cerebellar vermis

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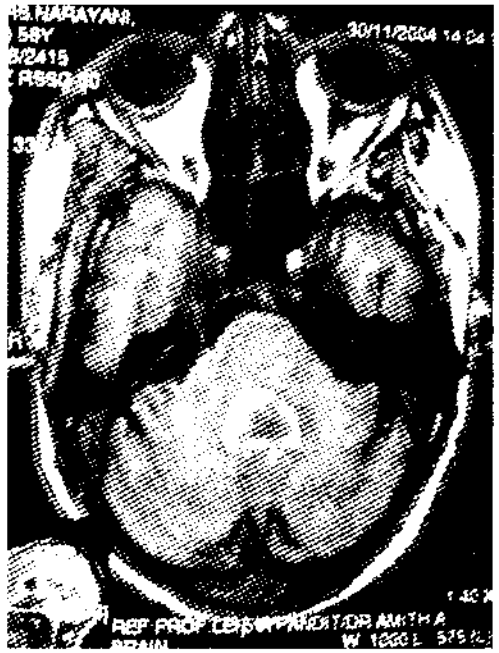

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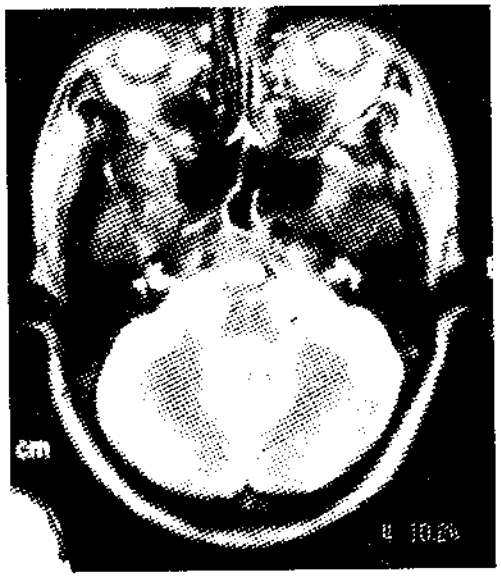


Fig. 7.5 MRI scan of the same patient showing haemorrhage involving cerebellar vermis (A. T1 image, B. T2 image)

Laboratory Investigations

Complete blood count
Platelet count
Bleeding time
Prothrombin and partial thromboplastin times
Liver and renal function tests

Lumbar puncture is contraindicated because it may precipitate a herniation syndrome in patients with a large haematoma.

Imaging

CT Scan

CT scan (without contrast) will confirm the site and size and it is superior to MRI for detecting intracranial haemorrhage of less than 48 hours duration. Characteristically infarction appears as low-density area on CT scan.

Cerebral Angiography

If the patient's condition permits further intervention, cerebral angiography may be undertaken thereafter to determine the presence of an aneurysm or arteriovenous malformation.

Treatment

Generally conservative and supportive.
Analgesics, tranquillizers, antiemetics, and laxatives.
Decompression is helpful, however, when a superficial haematoma in cerebral white matter is exerting a mass effect and causing incipient herniation.
In patients with cerebellar haemorrhage, prompt surgical evacuation of the haematoma will help to decompress the brain stem structures.
Treatment of underlying disease (control of blood pressure, clipping of aneurysm)
Chest physiotherapy and care of the airway will reduce the risk of pneumonia.
Nutritional support
Good nursing care to prevent bed sores
Early physiotherapy will reduce the risk of contractures, pain, and stiffness in hemiplegic limbs and leads naturally on to active physical rehabilitation.

Complications

Cerebral oedema
Transtentorial herniation
General complications of acute paralysis
Bronchopneumonia
Venous thromboembolism
Pressure sores
Septicaemia
Urinary infection
Contractures in spastic limbs; frozen shoulder

Prevention of Stroke

Primary prevention in asymptomatic individuals

Detection and adequate treatment of hypertension
Dietary means (more exercise, reduce weight, less salt, and reduce heavy alcohol consumption) and the reduction of cigarette-smoking
Reduce cholesterol levels

Secondary prevention in patients who have already experienced a transient ischaemic attack or stroke

Control of blood pressure

Phenol nerve block. In the stroke patient, phenol nerve blocks occasionally are indicated to change the paralysis from spastic to flaccid and thus make the extremity more responsive to corrective exercises, corrective posturing, or bracing. Open intraneural phenol nerve blocks can be expected to diminish muscle tone for about 6 months.

Functional electrical stimulation (FES). In functional electrical stimulation, function is restored in paralyzed muscles by electrical stimulation. The aim is to have functional muscle control occur during stimulation, but occasionally a carryover occurs and the muscle comes under voluntary control even during periods without electrical stimulation.

CHAPTER 8

Head Injury

Most of the head-injury victims are young adult males and head injury is a leading cause of mortality and morbidity. More than half of the more severe injuries are caused by road traffic accidents; other causes are falls, sports and assaults.

PATHOLOGY

Injury can involve one or more of the following:

a) Scalp – laceration, avulsion, contusion
b) Bone – different type of fractures
c) Membranes – extradural haematoma, subdural haematoma (acute/chronic)
d) Brain – contusion, laceration, intracerebral haematoma, diffuse axonal injury
e) Intracranial – intraventricular haemorrhage

SALIENT FEATURES

Scalp Injuries

The scalp is very vascular and laceration can cause severe loss of blood. The vessels within the scalp do not constrict when injured because the wall is adherent to the surrounding fiber fatty tissue in the subcutaneous area. Bleeding can be controlled by applying pressure or suturing the scalp. There will be excellent healing of the scalp wounds after surgical toilet and closure because of its generous blood supply.

Skull Injuries

Skull factures may occur with no associated neurological damage and conversely, fatal injury to membranes, blood vessels and the brain may

occur without overlying fracture. Skull fracture can be described in following ways:

Location – Vault, posterior fossa and base of the skull
Shape – Linear (fissure) or comminuted into fragments
Undisplaced and depressed
Open, or compound (i.e., associated with a local wound which creates a passage between the exterior and the interior of the cranial cavity)

Diffuse Axonal Injury (Figure 8.1)

Diffuse axonal injury results when the shear strains between different parts of the brain cause distortion, stretching, and even tearing of the axons in the white matter of the cerebral hemispheres and brain-stem. Clinically their characteristic feature is an immediate impairment of consciousness.

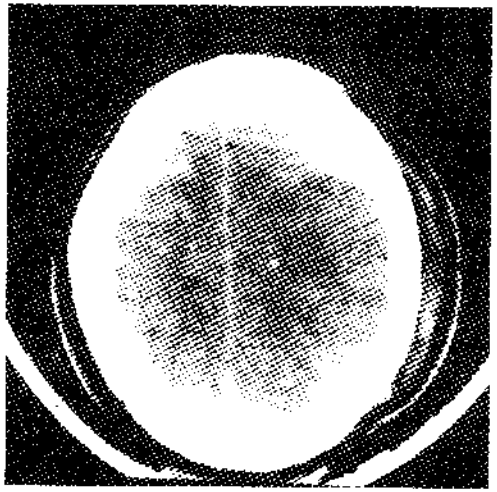


Fig. 8.1 CT scan showing multiple petechial haemorrhages

Contusions and Brain Lacerations (Figure 8.2)

These lesions are found focally, predominantly in the cerebral cortex, located maximally on the undersurfaces of the frontal and temporal lobes, and are due to displacement of the brain and contact with the sharp bony ridges at the base of the skull. When contusions occur in an eloquent area of the cortex there may be focal neurological signs in the acute stage. Contusions in the frontal and temporal lobes also may contribute to the changes in personality and mental state and the epilepsy that can follow a head injury.

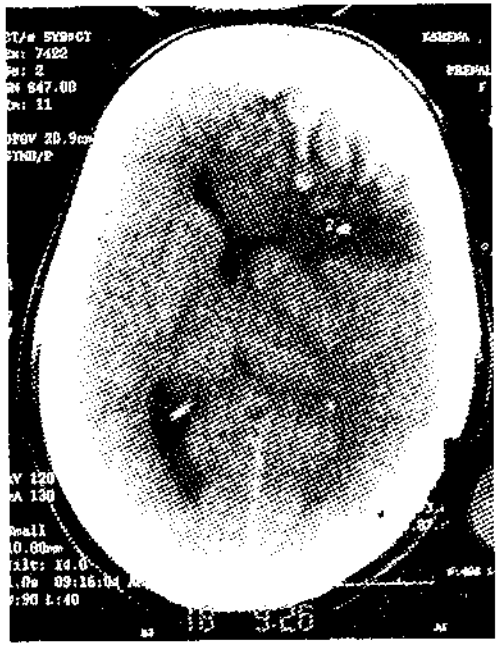


Fig. 8.2 CT scan showing left frontal contusion

Traumatic Intracranial Haematomas (Table 8.1)

Extradural Haematoma (Figure 8.3)

An extradural haematoma is usually associated with a skull fracture and is the result of bleeding between the site of the fracture and the underlying dura.

Subdural Haematoma (Figure 8.4)

In subdural haematoma there is accumulation of blood in subdural space (between dura mater and arachnoid). Subdural haematoma may be acute, subacute, and chronic. These lesions may be unilateral or bilateral.

Intracerebral Haematoma (Figure 8.5)

Intradural haematomas may be traumatic or spontaneous and these are due to disruption of cerebral vessels in brain parenchyma.

Cerebral Oedema

The damaged brain around a contusion or underneath a blood clot may become progressively swollen; this may involve one or both cerebral hemispheres.

Fig. 8.3 CT scan showing right temporo-parietal extradural haematoma

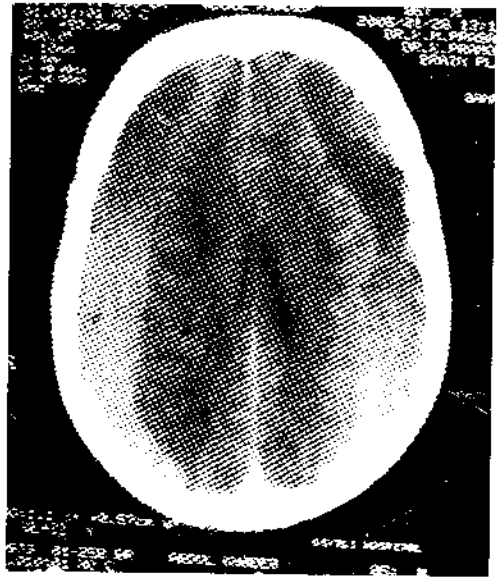


Fig. 8.4 CT scan showing bilateral chronic subdural haematoma

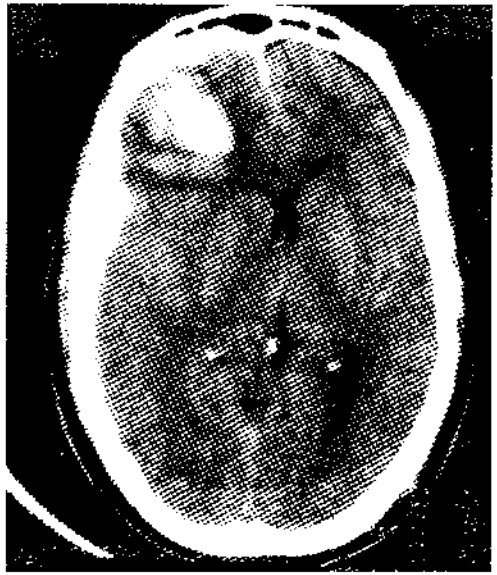


Fig. 8.5 CT scan showing right frontal intracerebral haematoma

Other Neurological Complications

Injury to the cranial nerves
(Olfactory, optic, occulomotor, facial, and auditory nerves)
Injury to the pituitary gland and hypothalamus
Deafness and dizziness due to damage to middle and inner ear

Table 8.1 Summary of intracranial lesions

Type	Salient features	Management
Concussion	Temporary neural dysfunction Recovery is usually complete within 24 to 48 hours CT scan–normal	
Epidural haematoma	Most common in 20 to 40 year olds. Most result from arterial bleeding. Blood commonly accumulates between skull and dura. Injury to middle meningeal artery CT will show collection of blood	Surgical evacuation

contd.

Contusion	Due to acceleration-deceleration or coup-contrecoup injuries Continous increase in size can lead to progressive neurologic deterioration	If large surgical evacuation Measures to reduce cerebral oedema
Skull fractures	May not be problematic unless brain is exposed or bone fragments are driven into neural tissue. Anterior fossa basilar (periorbital ecchymosis, anosmia), CSF rhinorrhea. Middle cranial fossa (CSF otorrhea, haemotympanium, facial paralysis)	Closed undisplaced fractures - conservative Open and depressed fractures–surgery

Radiological Investigations

Skull Radiograph

Skull fracture
Pneumocephalus

CT Scan

CT scanning can detect the presence of intracranial haematomas and skull fractures.

Management

The goal of emergency care of patients with head trauma is to recognize and treat life-threatening conditions and to eliminate or minimize the role of secondary brain injury. Patients with severe head trauma are at increased risk of developing cerebral oedema, respiratory failure, and herniation secondary to the increased ICP; therefore, frequent serial assessments of the neurologic status must be performed.

CHAPTER 9

Meningitis

Meningitis is an inflammation of the leptomeninges with infection of the cerebrospinal fluid within the subarachnoid space of the brain and spinal cord, and the ventricular system.

PATHOLOGY

The microorganism typically enters the CNS by many routes (Table 9.1). Microorganisms can be transmitted to an infant via the intrauterine environment. The invading organism triggers an inflammatory response in the meninges and raised intracranial pressure. This inflammation will also block the arachnoid villi, leading to obstruction to CSF outflow with resultant hydrocephalus.

Table 9.1 Causes of acute meningitis

Bacterial	S. pneumonia N. meningitidis H. influenzae Group B streptococci Listeria monocytogenes Gram-ve bacilli Staphylococci
Viral	Enterovirus HIV
Rickettsial	R. rickettsii Typhus
Spirochetal	Treponema pallidum Borrelia burgdorferi

contd

Protozoa	Naegleria fowleri
Medications	NSAIDs Azathioprine Carbamazepine
Miscellaneous	SLE

SOURCE OF INFECTION

Haematogenous
Pneumonia
Empyema
Osteomyelitis
Endocarditis

Contiguous Spread
Sinusitis
Otitis media
Encephalitis
Myelitis

Post-traumatic
Skull fracture
Penetrating head wound

Iatrogenic
Lumbar puncture
Ventricular shunting

CLINICAL FEATURES

Symptoms

Infection and inflammation
Fever, chills, and malaise resulting from raised ICP
Headache, vomiting, and rarely, papilloedema (inflammation and oedema of the optic nerve)

Signs of Meningeal Irritation

Papilloedema
Papilloedema is suggestive of cerebral oedema or an intracranial space-occupying lesion such as a cerebral abscess or a subdural or epidural collection of pus.

The optic fundi should be examined as a prelude to lumbar puncture.
Nuchal rigidity
Positive Brudzinski's and Kernig's signs
Exaggerated and symmetrical deep tendon reflexes
Opisthotonos (a spasm in which the back and extremities arch backward so that the body rests on the head and heels)

Other Features of Meningitis

Sinus arrhythmias from irritation of the nerves of the autonomic nervous system
Irritability from increasing ICP
Photophobia, diplopia, and other visual problems from cranial nerve irritation
Delirium, deep stupor, and coma from increased ICP and cerebral oedema.
Infants may show signs of infection, but most are simply fretful and refuse to eat. In an infant, vomiting can lead to dehydration, which prevents formation of a bulging fontanelle, an important sign of increased ICP.
As the illness progresses, twitching, seizures (in 30% of infants), or coma may develop.
Physical examination must exclude otitis media, sinusitis, mastoiditis, and nasopharyngeal and other possible sites of sepsis.

COMPLICATIONS

Increased ICP (Cerebral oedema)
Hydrocephalus
Cerebral infarction
Cranial nerve deficits including optic neuritis and deafness
Unilateral or bilateral sensory hearing loss
Encephalitis
Paresis or paralysis
Endocarditis
Brain abscess
Syndrome of inappropriate antidiuretic hormone (SIADH)
Seizures
Coma
Mental retardation–particularly in children

DIFFERENTIAL DIAGNOSIS

Meningeal irritation is seen in many acute febrile conditions, especially in children.
Local infections of the nasopharynx
Cervical lymph nodes

Tetanus
Subarachnoid haemorrhage
Tuberculous and cryptococcal and other fungal meningitides
Viral meningitis

DIAGNOSIS

Lumbar Puncture

Elevated CSF pressure
Cloudy CSF
High protein level
Positive Gram stain and culture
Decreased glucose concentration

Culture

Blood
Urine
Nose and throat secretions

Chest X-ray

Pneumonitis
Lung abscess
Tubercular lesions
Fungal infection

Sinus and Skull X-rays

Cranial osteomyelitis
Paranasal sinusitis
Skull fracture

White Blood Cell Count

Leucocytosis

Computed Tomography

Hydrocephalus or rule out cerebral haematoma, haemorrhage, or tumour. Should be done before performing lumbar puncture.

TREATMENT

Antibiotics

Usually, IV antibiotics for at least 2 weeks, followed by oral antibiotics. Selected by culture and sensitivity testing (Table 9.2)

Table 9.2 Empirical regimes of antibiotics in meningitis

Type of meningitis	Antibiotic regimes
Neonatal meningitis	Aminoglycoside + penicillin or ampicillin Third-generation cephalosporin + penicillin or ampicillin
Community aquired	Ceftriaxone, cefotaxime, or chloramphenicol plus ampicillin
Spontaneous meningitis	Penicillin alone Alternatively, cefotaxime or ceftriaxone
Post-traumatic meningitis	Cefotaxime, ceftriaxone, or chloramphenicol + ampicillin Ceftazidime, cefotaxime, or ceftriaxone
Shunt-associated meningitis	Vancomycin In acute presentation Cefotaxime, ceftazidime or ceftriaxone + vancomycin with removal of infected shunt

Once the aetiological agent has been isolated and its susceptibilities determined, the empirical treatment should be changed, if necessary, to an agent or agents specific for the isolate.

Mannitol

To control intracranial pressure

Anticonvulsant

To prevent or control seizure activity

Headache and Fever

Analgesics (Aspirin or acetaminophen)

Supportive Measures

Bed rest
Parenteral fluids
Treatment of underlying conditions (e.g., endocarditis or pneumonia, skull fracture etc)

PROGNOSIS AND SEQUELAE

Permanent neurological sequelae
Mental retardation, deafness and other cranial nerve deficits, and hydrocephalus.

CHAPTER 10

Encephalitis

Acute encephalitis represents inflammation of brain tissue and may occur concurrently with meningeal irritation ("meningoencephalitis").

AGENTS

Viral

Herpes simplex
Varicella-zoster virus
Epstein-Barr virus
Rubeola virus
Enteroviruses (Coxsackie virus, echovirus, and poliovirus) and togaviruses (equine encephalitis virus)

Nonviral (potentially treatable causes)

Rocky Mountain spotted fever
Malaria
Brucellosis
Amebic (Naegleria) infection
Syphilis
Lyme borreliosis
Toxoplasmosis
Measles and rabies vaccines

PATHOLOGY

The pathological effects of viral infections on the central nervous system include:
Destruction and phagocytosis of neurones (neuronophagia) as a result of either viral invasion per se or immune lyses.

Demyelination
Inflammatory oedema with the compressive effects of raised intracranial pressure; and, in some cases, vascular lesions

CLINICAL FEATURES

Fever
Headache
Altered state of consciousness
Focal neurological deficits
Seizures
Autonomic abnormalities
High incidence of permanent and disabling sequelae
Herpes simplex is the most common cause of sporadic, focal encephalitis with a mortality rate of 10% to 40%, and a corresponding high incidence of serious residual deficits. Herpes simplex encephalitis characteristically is associated with early behavioral disorders because the virus tends to localize in the temporal lobes. Seizures occur early in the course in about half of cases.
Myalgia is particularly severe with Coxsackie B infections.

INVESTIGATIONS

Evaluation of encephalitis requires a comprehensive history, physical examination, and LP.

Lumbar Puncture

Examination of the CSF usually reveals a lymphocytic pleocytosis and elevated protein, but indices may be normal early on. Culture of the CSF usually does not yield virus.

CT Scan

CT scan will show evidence of inflammation and oedema in the characteristic areas.

MRI

MRI will show evidence of inflammation and oedema in the characteristic areas.

EEG

EEG with periodic sharp wave activity temporally on a background of focal or diffuse slowing should prompt empiric treatment with intravenous acyclovir.

Brain Biopsy

Brain biopsy remains the gold standard of diagnosis.

PCR (Polymerase chain reaction)

PCR to detect herpes simplex viral DNA in CSF is now known to be highly sensitive and specific and should be considered the strategy of choice when herpes simplex encephalitis is considered.

DIFFERENTIAL DIAGNOSIS

Partially treated bacterial meningitis
Tuberculous meningitis
Fungal meningitis
Amoebic meningitis
Neoplastic meningitis
Granulomatous meningitis
Idiopathic meningitis

TREATMENT

Successful therapy depends on a high index of suspicion leading to early diagnosis.

Supportive Treatment

General care
Control of fever and maintenance of fluid and electrolyte balance

Cerebral Oedema

In viral encephalitis, the CSF pressure may be very high and focal areas of infection with oedema, vascular engorgement, or haemorrhage may act as a mass lesion. To prevent brain herniation osmotic agents (mannitol) may be used in acute situations, and high-dose steroids may be used.

Seizures

Seizures resulting from viral encephalitis may be extremely difficult to control and can be treated with appropriate anticonvulsants.

Specific Antiviral Therapy

Herpes simplex encephalitis
Acyclovir, 10.0 to 12.5 mg/kg IV q8h
The patient should be adequately hydrated, and each dose should be slowly infused over 60 minutes to prevent the drug from precipitating in the renal tubules. The optimum duration of therapy is unknown, but 10 to 14 days is common.

Toxicity. Acyclovir is generally well tolerated, but local irritation, nausea, and headache can occur with IV infusion. About 1% of patients receiving intravenous acyclovir develop encephalopathy, which can be difficult to distinguish from the underlying encephalitis. Abnormal bone marrow and hepatic function have been reported in immunocompromised patients.

CMV Infections

Ganciclovir (DHPG) has broad-spectrum antiviral activity against herpesviruses but has achieved widest clinical use in CMV infections. It has demonstrated efficacy in CMV retinitis and is used, although without documented effect, in CMV encephalitis and myelitis. Improvement of CMV polyradiculitis has been demonstrated in a few cases.

Dosage–Ganciclovir, 5.0 mg/kg IV q12h, is infused over 1 hour. The dosage must be adjusted in renal insufficiency. The total duration of therapy is generally 14 to 30 days, depending on the clinical response. In AIDS patients, maintenance therapy at a dosage of 5 mg/kg/day or 6 mg/kg/day, 5 days/week, may have to be administered for life to prevent relapse.

Toxicity–The toxicity is dose-related and is directed primarily at rapidly dividing cells. Inhibition of spermatogenesis, bone marrow suppression, and GI reactions are common. Encephalopathy is uncommon.

Foscarnet has a spectrum of antiviral activity similar to that of ganciclovir. It is effective in CMV retinitis and can be used as a second-line agent in patients with ganciclovir-resistant CMV causing nervous system disease.

Dosage is 60 mg/kg IV q8h for 14 days, infused over 1 hour by infusion pump only. In AIDS patients, maintenance therapy, 90 to 120 mg/kg/day IV, is recommended, administered as a 2-hour pump infusion. Oral ganciclovir has been successful as maintenance therapy in CMV retinitis and is a reasonable alternative.

Pharmacokinetics–Foscarnet is excreted by the kidney and has a plasma half-life of 3.3 hours. Levels in CSF reach 40% of those in plasma.

Toxicity–Renal toxicity is dose-limiting in up to 23% of patients. Derangements of calcium, potassium, phosphate, and magnesium levels can occur. The dosage must be adjusted in renal failure. Acute overdose can cause seizures or cardiac arrhythmias and is potentially fatal.

NEUROLOGICAL COMPLICATIONS

Mental retardation
Loss of memory
Speech abnormalities
Hemiparesis
Ataxia
Brain-stem and cranial nerve lesions
Recurrent convulsions

CHAPTER 11

Parkinson's Disease

Parkinsonism is a relatively common disorder that occurs in all ethnic groups, with an approximately equal sex distribution. The most common variety, idiopathic Parkinson's disease (paralysis agitâns), begins most often between 45 and 65 years of age. The disease is characterized by slowly progressive akinesia, rigidity, postural abnormality, and tremor.

AETIOLOGY

Idiopathic parkinsonism
Familial – mutations of several different genes (rare)
Postencephalitic parkinsonism
Exposure to certain toxins (e.g., manganese dust, carbon disulfide)
Severe carbon monoxide poisoning
Use of 1-methyl-4-phenyl-1,2,5,6-tetrahydropyridine (MPTP) for recreational purposes.
Drug-induced parkinsonism – reserpine, phenothiazines, metoclopramide and butyrophenones
Hemiparkinsonism may be the presenting feature of a progressive space-occupying lesion (extremely rare)

PATHOLOGY

The pathology in the substantia nigra consists of loss of pigmented neurones in the zona compacta, with eosinophilic inclusions, called Lewy bodies, in remaining neurones. This dopamine depletion due to degeneration of the dopaminergic nigrostriatal system leads to an imbalance of dopamine and acetylcholine, which are neurotransmitters normally present in the corpus striatum. Treatment is directed at redressing this imbalance by blocking the effect of acetylcholine with anticholinergic drugs or by the administration of levodopa, the precursor of dopamine.

CARDINAL FEATURES

Following are the cardinal features of parkinsonism and may be present in any combination. There is typically no muscle weakness and no alteration in the tendon reflexes or plantar responses.

Resting Tremors

Parkinsonian tremors (four to six cycles per second) is present at rest, is decreased by action, is increased by emotion or stress, and disappears during sleep. The arms are most often affected initially, but tremor may spread to involve the face, jaw, and legs.

Rigidity

Rigidity (an increase in resistance to passive movement) is responsible for the characteristically flexed posture seen in many patients. Rigidity affects all muscles, but is most marked in the neck and trunk, and in proximal muscles at the shoulder or hip. When tremor coexists, the smooth plastic nature of rigidity may be broken up by rhythmic catches (cogwheel phenomenon).

Bradykinesia

The most disabling symptoms of parkinsonism are due to bradykinesia, manifested as a slowness of voluntary movement and a reduction in automatic movements such as swinging of the arms while walking. Curiously, however, effective voluntary activity may briefly be regained during an emergency (e.g., the patient is able to leap aside to avoid an oncoming motor vehicle).

Postural Instability

It is difficult for the patient to arise from a sitting position and begin walking. The gait is characterized by small shuffling steps and a loss of the normal automatic arm swing; there may be unsteadiness on turning, difficulty in stopping, and a tendency to fall.

Box 11.1 Other associated features

- Mild decline in intellectual function
- Relatively immobile face
- Widened palpebral fissures
- Infrequent blinking
- Fixed of facial expression
- Seborrhea of the scalp and face
- Drooling of saliva - due to impairment of swallowing
- Soft and poorly modulated voice

contd.

- Variable rest tremor and rigidity in some or all of the limbs
- Slowness of voluntary movements
- Impairment of fine or rapidly alternating movements
- Micrographia

DIFFERENTIAL DIAGNOSIS

Depression (expressionless face, poorly modulated voice, and reduction in voluntary activity)
Wilson's disease
Huntington's disease
Shy-Drager syndrome
Progressive supranuclear palsy
Creutzfeldt-Jakob disease
Cortical-basal ganglionic degeneration

TREATMENT

Table 11.1 Drugs

Drug	Indications	Side effects
Amantadine	Patients with mild symptoms No disability Improves all of the clinical features of parkinsonism	Restlessness, confusion, depression, skin rashes, oedema, nausea, constipation, anorexia, postural hypotension, and disturbances of cardiac rhythm
Anticholinergic drugs	More helpful in alleviating tremor and rigidity Less effective in bradykinesia	Dryness of the mouth, nausea, constipation, palpitations, cardiac arrhythmias, urinary retention, confusion, agitation, restlessness, drowsiness, mydriasis, increased intraocular pressure, and defective accommodation
Levodopa	Improves all of the major features of parkinsonism, including bradykinesia Does not stop progression	Nausea, vomiting, and hypotension, but cardiac arrhythmias, dyskinesias, restlessness, confusion, and other behavioural changes

contd.

Dopamine agonists (bromocriptine and pergolide)	Patients refractory to levodopa	Anorexia, nausea, vomiting, constipation, postural hypotension, digital vasospasm, cardiac arrhythmias, various dyskinesias and mental disturbances, headache, nasal congestion, erythromelalgia, and pulmonary infiltrates

Surgical Measures

Thalamotomy or pallidotomy – May be helpful for patients who become unresponsive to medical treatment or have intolerable side effects from antiparkinsonian agents.
Surgical implantation of adrenal medullary or fetal substantia nigra tissue into the caudate nucleus has been reported with some benefit but still under evaluation.

Brain Stimulation

High-frequency thalamic stimulation – effective in suppressing the rest tremor of Parkinson's disease.
Chronic bilateral stimulation of the subthalamic nuclei or globus pallidus internus – may benefit all the major features of the disease.

Physical Therapy

Parkinson's disease produces a wide range of functional locomotor disabilities that can be assessed and managed by the physiotherapist and occupational therapist (Box 11.2).

Box 11.2

Eating
Washing
Dressing
Walking
Training to maintain equilibrium
To restore effective gait patterns

The quality of life can often be improved by the provision of simple aids to daily living, e.g., rails or banisters placed strategically about the home, special table cutlery with large handles, nonslip rubber table mats, and devices to amplify the voice.

CHAPTER 12

Multiple Sclerosis

SYNONYM – DISSEMINATED SCLEROSIS

This common neurologic disorder, which probably has an autoimmune basis, has its greatest incidence in young adults.
No one triggering agent has been identified
More likely to occur following viral exposure (measles, mumps, rubella, and Epstein-Barr virus infection)
Genetic susceptibility – likely (based on twin studies, familial cases, and an association with specific HLA-DR2 antigen).

PATHOPHYSIOLOGY

Myelin injury blocks saltatory conduction through myelinated axons.
Partially demyelinated pathways cannot transmit fast trains of impulse.
Reduced velocity
Delay in arrival of evoked potentials.

Partially demyelinated axons discharge spontaneously
Unpleasant distortions of sensation and facial myokymia
Increased mechanical sensitivity results in movement-induced phosphenes and the electric sensation provoked by neck flexion (Lhermitte's symptom)
Increased temperature sensitivity
Paroxysmal symptoms of demyelination such as trigeminal neuralgia or tonic brain-stem seizures.

CLINICAL FEATURES

Symptoms may disappear after a few days or weeks, although examination often reveals a residual deficit. The common clinical features include:
Weakness

Numbness
Tingling
Unsteadiness in a limb
Spastic paraparesis
Retrobulbar neuritis
Diplopia
Disequilibrium
Sphincter disturbance (urinary urgency or hesitancy)
Constipation
There is an interval of months or years after the initial episode before new symptoms develop or the original ones recur (relapsing-remitting disease). Eventually, however, relapses and usually incomplete remissions lead to increasing disability, with weakness, spasticity, and ataxia of the limbs, impaired vision, and urinary incontinence.

Neurological Examination

One or more deficits listed below.
Optic atrophy
Nystagmus
Dysarthria
Pyramidal type of weakness
Sensory deficits
Cerebellar dysfunction
Internuclear ophthalmoplegia
These deficits may involve some or all of the limbs.
Less commonly, symptoms are steadily progressive from their onset, and disability develops at a relatively early stage (primary progressive disease).
A number of factors (e.g., infection, trauma) may precipitate or trigger exacerbations.
Relapses are also more likely during the 2 or 3 months following pregnancy, possibly because of the increased demands and stresses that occur in the postpartum period.

LABORATORY INVESTIGATIONS

CSF

Cerebrospinal fluid analysis provides information, which is complementary to imaging abnormalities. The cell count rarely exceeds 50 lymphocytes/mm^3, and is normal in more than 50 per cent of patients. There is a rise in total protein with a specific increase in the immunoglobulin concentration and the presence of oligoclonal bands on protein electrophoresis. The presence of such bands is not specific, however, since they have been found in a variety of inflammatory neurologic disorders and occasionally in patients with vascular or neoplastic disorders of the nervous system.

Electrophysiology

Demyelination can be detected in clinically unaffected pathways using visual, auditory, somatosensory, central motor and event-related potentials. In multiple sclerosis their latencies are characteristically delayed, whereas, except in acute lesions, the amplitude is unaffected. Evoked potentials add little in situations where the pathway under investigation is clinically affected and their role as an adjunct to diagnosis has largely been replaced by imaging techniques.

IMAGING

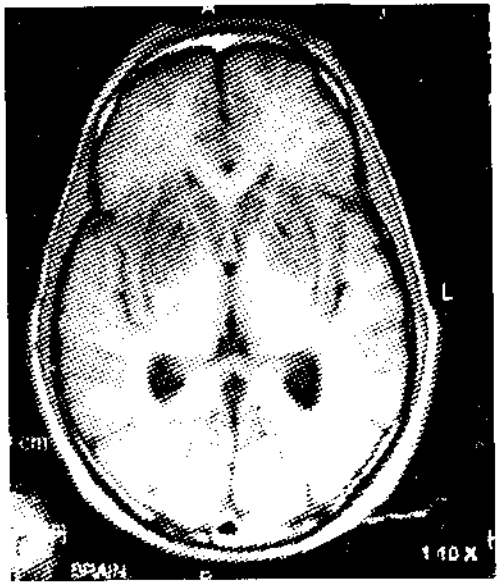
CT Scan

Low-density lesions, corresponding to areas of demyelination can be demonstrated on CT scan, however the CT scan is less helpful.

MRI

MRI of the brain or cervical cord will show the lesions and will also demonstrate the presence of a multiplicity of lesions (Figure 12.1A-E). MRI will also exclude a congenital or acquired surgically treatable lesion (e.g., Arnold-Chiari malformation, tumours). MRI will show white-matter abnormalities, each corresponding to areas of histological damage. These lesions will enhance after contrast (Gadolinium) administration.

A



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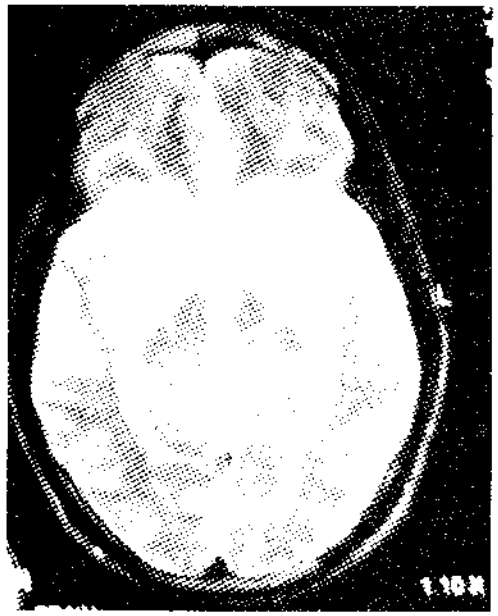

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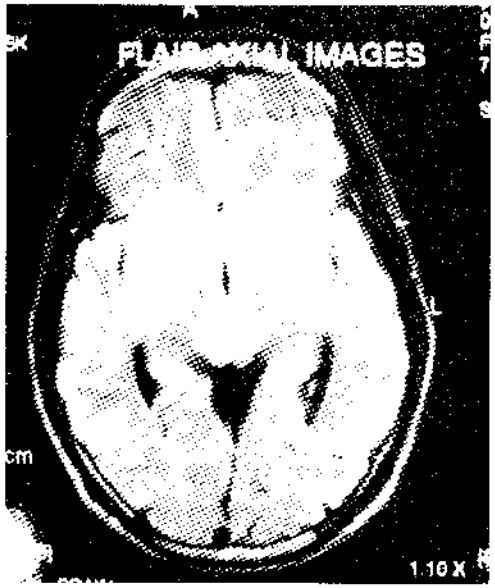

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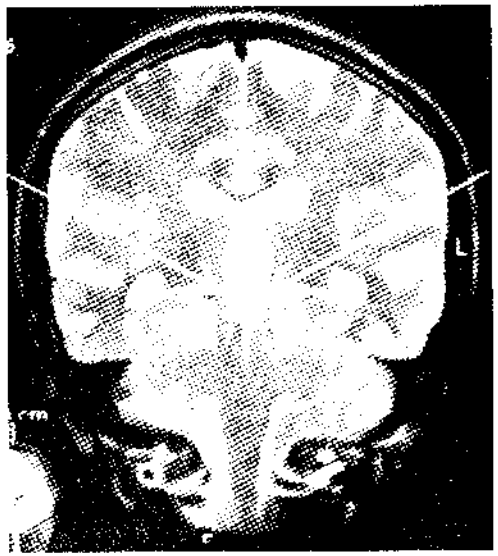

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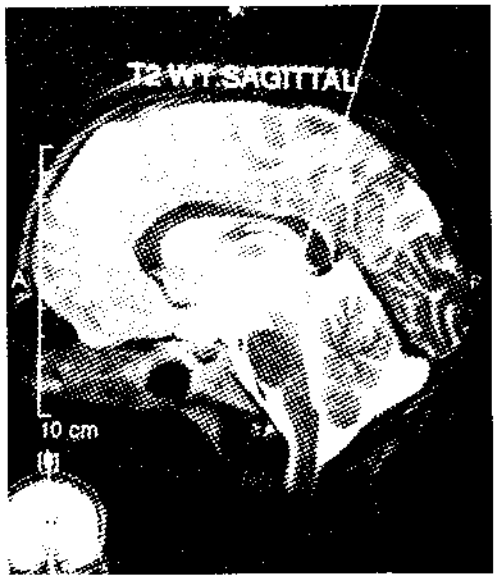


Fig. 12.1 MRI images of a patient with multiple sclerosis
(A) T1 axial image (B) T2 axial image
(C) FLAIR image (D) T2 coronal image
(E) T2 sagittal image

DIAGNOSIS

Multiple sclerosis should not be diagnosed unless there is evidence that two or more different regions of the central white matter have been affected at different times. A diagnosis of clinically definite disease can be made in patients with a relapsing-remitting course and evidence on examination of at least two lesions involving different regions of the central white matter.

DIFFERENTIAL DIAGNOSIS

Progressive spinal disease with a structural lesion (i.e., tumour)
Lesions at the foramen magnum
Brain-stem or spinal arteriovenous malformations
Hereditary spastic paraplegia
Systemic lupus erythematosus
Sarcoidosis
Tuberculous meningitis
Subacute combined degeneration of the spinal cord

TREATMENT

At least partial recovery from acute exacerbations can reasonably be expected, but further relapses may occur without warning, and there is no means of preventing progression of the disorder.

Acute Relapse

Recovery from acute relapses may be hastened by treatment with corticosteroids. Initially methylprednisolone, 1 g intravenously for 3 days is given followed by a high dose (e.g., prednisone, 60 or 80 mg) is given daily for 1 week, after which medication is tapered over the following 2 or 3 weeks. Long-term treatment with steroids provides no benefit and does not prevent further relapses.

Relapsing-remitting Disease

In patients with relapsing-remitting or secondary progressive disease, many drugs have been tried with variable results. These include beta interferon, cyclophosphamide, azathioprine, methotrexate, cladribine, or mitoxantrone. There is little evidence that plasmapheresis enhances any beneficial effects of immunosuppression in multiple sclerosis.

Intravenous immunoglobulins may reduce the clinical attack rate in relapsing-remitting disease, but the available studies are inadequate to permit treatment recommendations.

Supportive Measures

Treatment of Spasticity (See Chapter 42)

Neurogenic Bladder

Mild urinary symptoms – Abdominal pressure or perineal stimulation
Mild incontinence, urgency or frequency – respond well to drugs(oxybutynin or propantheline)
Severe detrusor and sphincter dysfunction – intermittent self-catheterization
Permanent use of a catheter is preferable to dribbling incontinence, with its attendant risks of skin excoriation.
Avoid excessive fatigue.
Bed rest during acute relapse.
Constipation should be managed by dietary alteration and the use of bulk laxatives, avoiding agents that act directly on the bowel wall.

PROGNOSIS

The prognosis is relatively good when sensory or visual symptoms dominate the illness.
Motor involvement, especially when co-ordination or balance are disturbed, has a less good prognosis.
The outlook is also poor in older-onset patients and these are often males.
Frequent, prolonged relapses with incomplete recovery and a short interval between the initial episode and first relapse carry a worse prognosis, but the main determinant of disability is onset of the progressive phase.
The risk of relapse is increased in the puerperium.

CHAPTER 13

Brain Tumours

Intracranial tumours may be benign or malignant, primary or secondary and the clinical presentation management of tumours within the cranial cavity is influenced as much by their exact location and size.

INCIDENCE AND DISTRIBUTION

Intracranial tumours – 8 per cent of all primary neoplasms
In children the central nervous system is the second most common site for primary tumours.
About 70 per cent of childhood intracranial neoplasms are infratentorial.
Intracranial tumours represent the sixth most common form of neoplasm in adults, of which 70 per cent occur above the tentorium.

AETIOLOGY AND ASSOCIATED LESIONS

Genetic factors – Von Recklinghausen's disease (neurofibromatosis) is associated with optic and hypothalamic glioma, schwannomas of various cranial nerves, most commonly the eighth, and intracranial meningiomas.
Tuberose sclerosis is associated with periventricular glioma, and von Hippel-Lindau disease with cerebellar haemangioblastoma which have a tendency to recur.
Immunosuppressive therapy – intracranial lymphoreticular neoplasms
High-dose irradiation – local meningiosarcoma or glioma following local irradiation of the scalp for ringworm or for pituitary neoplasms.

PATHOLOGY

Primary intracranial tumours may be derived from the skull itself, from any of the structures lying within it, or from their tissue precursors. They may be divided into tumours of neurectodermal origin (derived from the neural tube, neural crest, and ectoderm) and those of other cell types (Table 13.1)

Table 13.1 Histological types of brain tumours

Lowest grade gliomas	Pilocytic astrocytoma Subependymal giant cell astrocytoma Protoplasmic astrocytoma Ganglioglioma Xanthomatous astrocytoma Subependymoma
Lower grade gliomas	Fibrillary (gemistocytic, protoplasmic) Astrocytoma Ependymoma Oligodendroglioma Mixed oligoastrocytoma Optic nerve glioma
Higher-grade gliomas	Anaplastic astrocytoma Anaplastic oligodendroglioma Anaplastic mixed glioma
Meningioma	Benign Atypical Malignant
Primitive neuroectodermal tumours (PNET)	Medulloblastoma Ependymoblastoma Pineoblastoma
Pituitary tumours	Pituitary adenoma Pituitary carcinoma
Pineal tumours	Pineal cyst Pineocytoma Pineoblastoma
Choroid plexus tumours	Choroid plexus papilloma Choroid plexus carcinoma
Nerves and/or nerve sheath tumours	Neuroma Schwannoma Neurofibroma
Cysts	Arachnoid cysts Dermoid Epidermoid
Others	Craniopharyngioma Metastatic brain tumours

CLINICAL FEATURES

Cerebral neoplasms produce subacute and progressive neurologic signs and symptoms. The patient's symptoms are determined by the size, location, and rate of growth of the tumour, as well as the degree of peritumour cerebral oedema.

Raised Intracranial Pressure

Headache and Vomiting

The headaches may be mild, and are often bilateral or diffuse, without localizing value. The "early morning headache" and nausea usually thought to be associated with increased intracranial pressure is a pattern seen in only a small proportion of cases.

Focal Deficits

Focal deficits depend on the location of the lesion and the extent of the surrounding brain oedema.

Seizures

Generalized convulsions or focal seizures (or both) occur in about one third of patients with tumours in the supratentorial compartment. Seizures are more likely to accompany slowly growing tumours than highly malignant ones.

Haemorrhage

Haemorrhage into a highly vascular tumour can produce a sudden change in neurologic status that can be mistaken for a stroke syndrome. Glioblastoma multiforme and brain metastases from choriocarcinoma, melanoma, and anaplastic lung cancer are the tumours most likely to bleed spontaneously.

Frontal Lobe Lesions

Progressive intellectual decline
Slowing of mental activity
Personality changes
Contralateral grasp reflexes
Expressive aphasia (posterior part of the left inferior frontal gyrus)
Anosmia (pressure on the olfactory nerve)
Focal motor seizures or contralateral pyramidal deficits (precentral lesions)

Temporal Lobe Lesions

Seizures with olfactory or gustatory hallucinations

Licking or smacking of the lips
Impairment of external awareness
Depersonalization
Emotional changes
Behavioral disturbances
Sensations of deja vu or jamais vu
Micropsia or macropsia (objects appear smaller or larger than they are)
Visual field defects (crossed upper quadrantanopia)
Auditory illusions or hallucinations

Parietal Lobe Lesions

Contralateral disturbances of sensation
Sensory seizures
Cortical type of sensory loss or inattention (astereognosis)
Contralateral hyperpathia and spontaneous pain (thalamic syndrome)
Contralateral homonymous field defect
Lesions of the left angular gyrus cause Gerstmann's syndrome (a combination of alexia, agraphia, acalculia, right-left confusion, and finger agnosia)
Involvement of the left submarginal gyrus causes ideational apraxia

Occipital Lobe Lesions

Crossed homonymous hemianopia
Partial field defects
Visual agnosia both for objects and for colours
Visual hallucinations
Bilateral occipital lobe involvement—cortical blindness
Loss of colour perception
Prosopagnosia (inability to identify a familiar face)
Simultagnosia (inability to integrate and interpret a composite scene as opposed to its individual elements)

Brain Stem Lesions

Cranial nerve palsies
Ataxia
Incoordination
Nystagmus
Pyramidal and sensory deficits

Cerebellar Lesions

Marked ataxia of the trunk if the vermis cerebelli is involved
Ipsilateral appendicular deficits (ataxia, incoordination and hypotonia of the limbs) if the cerebellar hemispheres are affected.

DIFFERENTIAL DIAGNOSIS

Chronic subdural haematoma
Cerebral infarction
Cortical venous thrombosis
Benign intracranial hypertension
Cerebral abscess
Tuberculoma
Hydrocephalus

INVESTIGATIONS

Non-radiological Investigations

Visual fields charting - in cases of suspected optic nerve or chiasmal compression
Caloric testing, audiometry, and measurement of brain-stem evoked potentials in cases of possible acoustic tumour.
Endocrine assessment for cases of suspected pituitary adenoma.

Electroencephalogram

The electroencephalogram provides supporting information concerning cerebral function and may show either a focal disturbance due to the neoplasm or a more diffuse change reflecting altered mental status.

Lumbar Puncture

Lumbar puncture is rarely necessary; the findings are seldom diagnostic, and the procedure carries the risk of causing a herniation syndrome.

Chest Radiography

Suspected intracranial tumour to exclude primary neoplasm or tuberculosis.

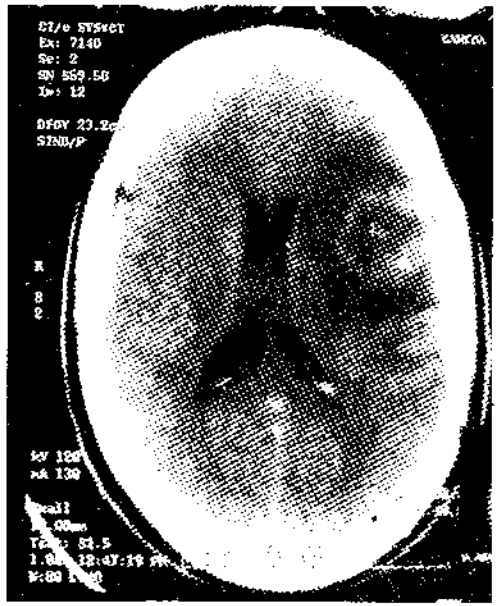
Skull Radiographs

Plain radiographs of the skull may show evidence of raised intracranial pressure in both adults and children (erosion of the dorsum sellae, copper beating of the skull vault, and separation of the sutures).
Calcification - oligodendrogliomas, craniopharyngioma, pineal tumour.
Hyperostosis or bony erosion - meningiomas, acoustic schwannomas.

CT Scan (Figure 13.1 a,b and 13.2 a,b)

CT scan without and with contrast reveals most brain tumours, and allows assessment of accompanying cerebral oedema, midline shift, and ventricular compression or obstructive hydrocephalus.

A



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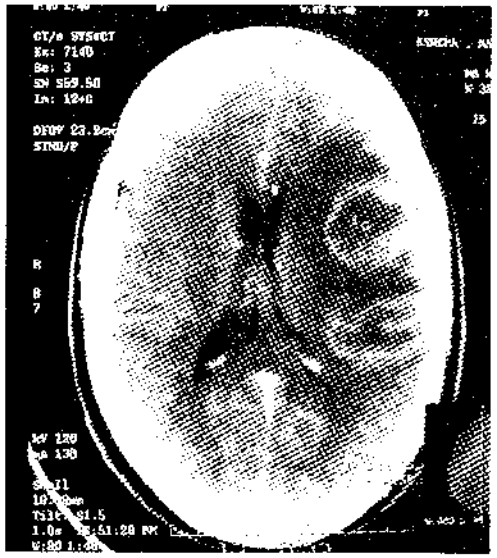


Fig. 13.1 CT scan of a patient with left temporal glioma (A) Plain study (B) Contrast study

A

B

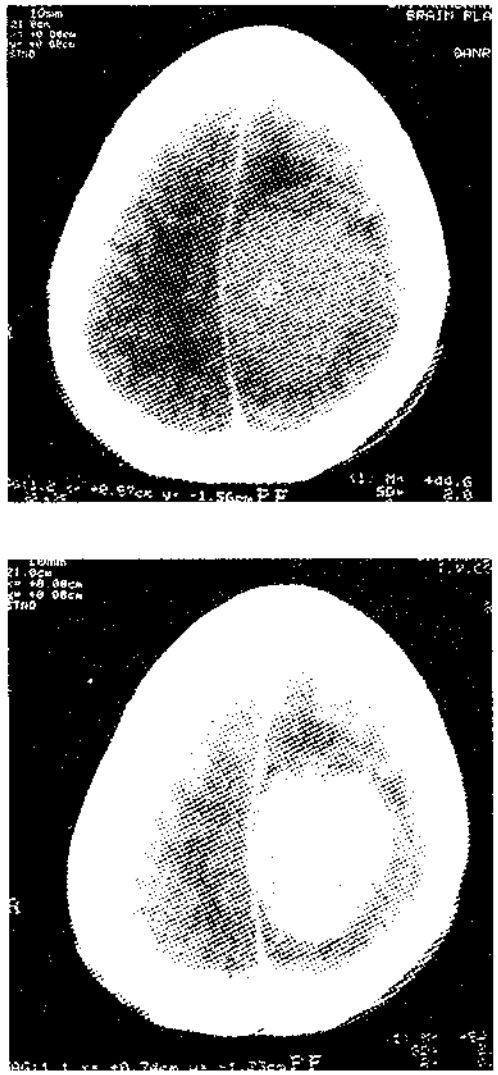


Fig. 13.2 CT scan of a patient with meningioma
(A) Plain study (B) Contrast study

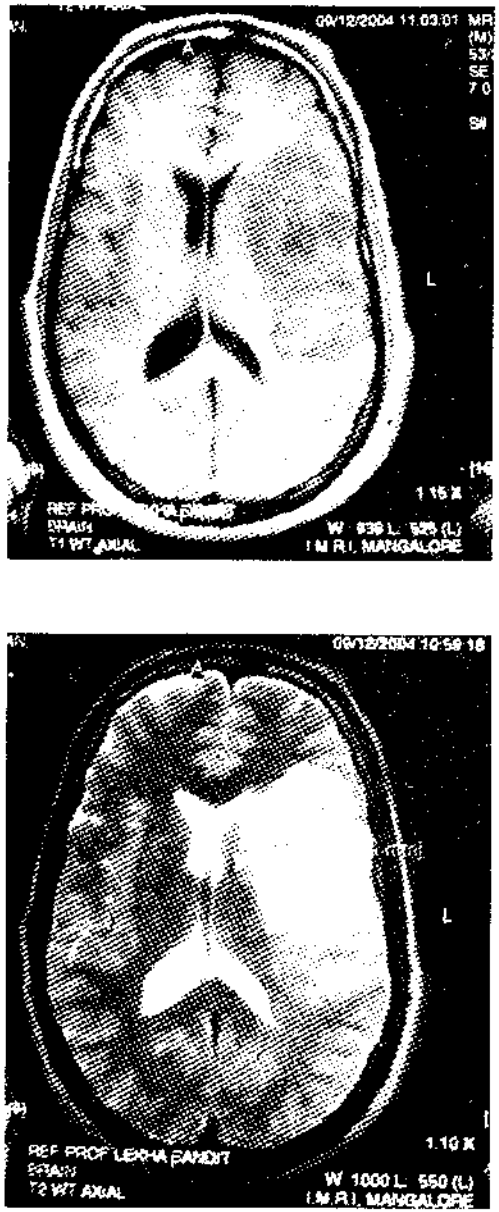
Magnetic Resonance Imaging (Figure 13.3 and 13.4)

Magnetic resonance imaging (MRI) without and with a contrast agent (gadolinium) is the most sensitive diagnostic study for detecting intracranial mass lesions. MRI is particularly useful for visualizing the brainstem and other posterior fossa structures that are not well seen on computed tomography (CT). The contrast agent helps differentiate between the

borders of the tumour and surrounding oedema, and facilitates detection of small lesions that might otherwise be missed.

A

B



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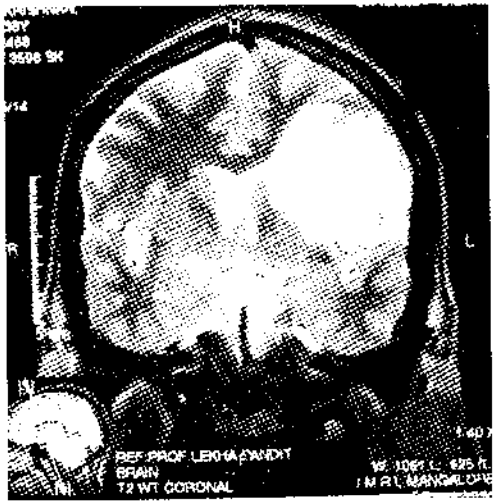


Fig. 13.3 MRI of a patient with glioma
(A) T1 axial image, (B) T2 axial image, (C) T2 sagittal image

Cerebral Angiography

Angiography may show stretching or displacement of normal cerebral vessels by the tumour and the presence of tumour vascularity. The presence of an avascular mass is a nonspecific finding that could be due to tumour, haematoma, abscess, or any space-occupying lesion.

A

B

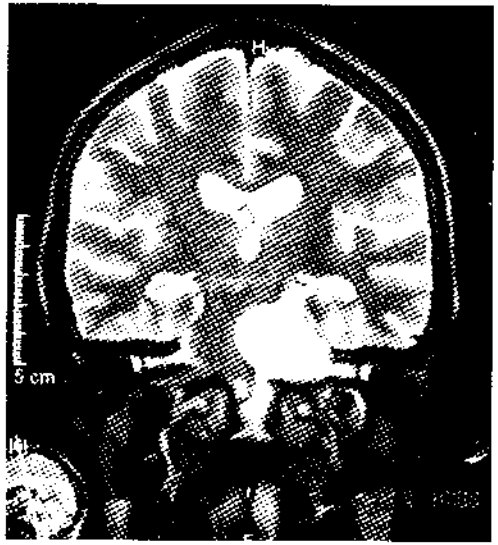


Fig. 13.4 MRI of a patient with left acoustic schwannoma
(A) T1 axial image, (B) T2 coronal image

MANAGEMENT

Management of Cerebral Oedema

Corticosteroids

High-dose corticosteroids can reduce tumour-associated cerebral oedema, thereby lowering intracranial pressure and palliating focal deficits. Dexamethasone (4 mg, six-hourly) has been the most widely used steroid for treating cerebral oedema, due to its relatively low mineralocorticoid (salt-retaining) side effects.

Frusemide or mannitol also can be used to decrease cerebral oedema.

Surgical Therapy of Intracranial Tumours

Histologic Confirmation

Direct histologic confirmation provides the basis for determining the prognosis and need for radiotherapy or chemotherapy.

Restoration of Impaired Neurologic Function

If the preoperative neurologic deficit is due to direct infiltration and destruction of brain tissue by tumour, surgical resection cannot be expected to improve the situation. Conversely, to the extent that the deficits are linked to pressure-related phenomena, they may be surgically reversible.

Surgical resection can be palliative, even in cases in which total resection is not possible, by providing an "internal decompression." Patients with benign brain tumours that cannot be totally resected, may be given many additional years of productive life by repeated decompressions at appropriate intervals.

Possibility of Cure

The complete removal of the brain tumour and permanent prevention of tumour recurrence can be achieved in many extra-axial tumours (e.g., meningiomas, schwannomas, pituitary adenomas). Very few intra-axial tumours can be resected completely or cured surgically.

Seizures

All patients with a history of seizures prior to surgery should be maintained on a proper anticonvulsant regimen (e.g., phenytoin or carbamazepine) during and after surgery.

PROGNOSIS

Glioblastomas have a mean survival time of only 6 to 12 weeks from presentation. Patients with low-grade tumours have a mean survival of about 9 months following surgery and radiotherapy.
Although there have been reports of up to 50 per cent 5-year survival in grade I cases.

Meningiomas, apparently completely resected, are still associated with a 5 to 10 per cent recurrence rate within 10 years, and this rises to over 20 per cent symptomatic recurrence, if fragments of tumour are known to have been left behind.

Cerebellar astrocytomas and haemangioblastomas are associated with a good prognosis after complete removal, although occasionally cerebellar astrocytoma may act in a more malignant fashion.

Fourth ventricular ependymoma has a poor prognosis if incompletely removed; the prognosis of medulloblastoma has already been described. The generally accepted recurrence rate of craniopharyngioma following radical surgery and irradiation is about 25 per cent within 5 years, but some reports suggest a less than 10 per cent recurrence rate.

Acoustic schwannomas will recur if incompletely removed, but their growth rate is usually slow and in a few elderly patients with large tumours, it may be preferable to accept this risk of slow recurrence and opt for a subcapsular resection. In all other instances radical removal should be attempted.

CHAPTER 14

Cerebral Palsy

Cerebral palsy is defined as a disorder of movement and posture caused by a nonprogressive defect or lesion in an immature brain.

The aim of treatment of children and adults with cerebral palsy is to improve function and prevent the development of deformities by physiotherapy, prescribing braces, and by reestablishing the balance of muscle forces through soft tissue releases, transfers, or bony reconstructions.

ETIOLOGY

The lesion responsible for cerebral palsy may have its origin in the prenatal, natal, or postnatal period (Table 14.1). Clinical type depends on the site of lesion in the brain (Table 14.2).

Table 14.1 Causes of cerebral palsy

Prenatal causes	Intrauterine infections TORCH (toxoplasmosis, rubella, cytomegalovirus, and herpes) Anoxia in the prenatal period Ruptured placenta Placental infarction Maternal pneumonia Cardiorespiratory disease Erythroblastosis fetalis Chemical or alcohol dependency in a mother Congenital brain defect (such as an absence of the corpus callosum)

contd.

Natal causes	Trauma or asphyxia occurring during labor Oxytocin augmentation Cord prolapse Breech presentation Prematurity Low birth weight (less than 2268 g) Periventricular haemorrhages
Postnatal causes	Encephalitis and meningitis Traumatic head injuries Anoxic encephalopathy from near-drowning Removal of brain tumours can result in a motor deficit

Table 14.2 Clinical types of cerebral palsy and sites of lesion

Clinical type	Site of lesion	Incidence
Spastic	Cerebral cortex	70–80%
Ataxic	Cerebellum	< 5%
Athetoid	Midbrain or base of the brain	10–15%
Mixed	Widespread brain involvement	10%
Hypotonic and rigid		Uncommon

CLINICAL FEATURES

Cerebral palsy is a nonprogressive neurological lesion. The brain lesion that causes the deficit in voluntary muscle control and posture, balance abnormalities, and disorders in tone remains constant. Clinical features of cerbral palsy can be calssifeid according to anatomical type or according to clinical presentation (Table 14.3 and Table 14.4).

TREATMENT

Though the brain lesion in the cerebral palsy is constant the natural history of this disease is not static. Growth and maturation of the child will often result in changing musculoskeletal problems. The goals of treatment may vary depending on the physiological and anatomic type of cerebral palsy and the amount of neurological involvement. Treatment goals should be individualized and be consistent with the expected benefits (Box 14.1).

Table 14.3 Anatomical classification of cerebral palsy and salient features

Anatomical type	Salient features
Monoplegia	Only one extremity is affected Either upper or lower Extremely uncommon type of paresis. Usually is seen after meningitis
Hemiplegia	Both extremities on the same side are affected Upper extremity typically is more affected than the lower one Results from bleeding in the middle cerebral artery involving the central portion of one hemisphere Associated with sensory disorders in the involved limbs
Paraplegia	Lower extremities are involved In true paraplegia upper extremity gross and fine motor controls are completely normal True spastic paraplegia is very rare MRI of spine to rule out spinal tumours and diastematomyelia
Diplegia	More often seen in premature infants Due to the loss of blood flow in the region of the periventricular areas Both lower extremities are always involved to a greater extent than the upper extremities Intelligence is usually normal
Quadriplegia	All four extremities are involved equally Typically, the patient has neck and trunk control.
Double hemiplegia	Due to intraventricular bleeding in the middle cerebral artery in both hemispheres of the brain Upper extremities are more involved than the lower extremities
Total body involvement	All extremities are involved, as well as the trunk and neck muscles Usually have problems with drooling, dysarthria, and dysphagia These patients require total care Cannot assist at all in the activities of daily living

Table 14.4 Clinical types of cerebral palsy and salient features

Clinical type	Signs and symptoms
Spastic	Most common type Due to impairment of the pyramidal tract Hyperactive deep tendon reflexes Increased stretch reflexes Rapid alternating muscle contraction and relaxation Muscle weakness Underdevelopment of affected limbs Muscle contraction in response to manipulation Tendency towards contractures Typical walking on toes with a scissors gait, crossing one foot in front of the other Most orthopaedic surgical procedures are designed for patients with spasticity.
Athetoid	Second most common clinical type Due to impairment of the extrapyramidal tract Involuntary movements usually affecting arms more severely than legs (e.g., grimacing, worm-like writhing, dystonia, sharp jerks) Difficulty with speech due to involuntary facial movements Increasing severity of movements during stress; decreased with relaxation and disappearing entirely during sleep
Ataxic	Due to impairment of the extrapyramidal tract Disturbed balance Incoordination (especially of the arms) Hypoactive reflexes Nystagmus Muscle weakness Tremor Lack of leg movement during infancy Wide gait as the child begins to walk Sudden or fine movements, impossible (due to ataxia)
Mixed	Spasticity and athetoid movements Ataxic and athetoid movements (resulting in severe impairment)

Box 14.1

To increase the patient's abilities as much as possible
To minimize impairments or disabilities
To increase the patient's:

- Emotional maturity
- Physical independence
- Cognitive abilities
- Speech or communication
- Socioeconomic independence
- Education and communication

To improve the activities of daily living, mobility, and ambulation.

For a patient with severe spastic quadriplegia, improving nursing care, hygiene, and sitting balance in a wheelchair should be the primary objective. In a patient with less severe spastic hemiplegia, the treatment goals may be to improve gait and make ambulation more energy efficient.

Treatment of Spasticity (See details in Chapter 42)

Longitudinal myelotomy
Stereotactic cortex ablation
Procedures on the deep cerebellar nuclei or the thalamus for extrapyramidal lesions where motion disorders originate
Electrical stimulation of the cerebellum
Electrical stimulation over the dorsal spinal cord
Transcutaneous electric stimulation
Phenol injection
Botulinum toxin
Baclofen
Selective dorsal rhizotomy

Indications for Reconstructive Surgery (See Chapter 49)

Surgery is most often indicated in patients with spastic cerebral palsy, less often in those with dyskinesia and athetosis.

To help correct local physical defects that interfere with the patient's rehabilitation.

To help to correct defects that interfere with nursing care.

To release contractures (discussed later)—equinus deformities of the ankles and flexion contractures of the knees and hips.

CHAPTER 15

Hydrocephalus

Hydrocephalus is broadly defined as a disturbance of formation, flow, or absorption of cerebrospinal fluid that leads to an increase in volume occupied by this fluid, in the central nervous system.

NORMAL CSF PATHWAY

Normal route of CSF from production to clearance is the following: From the choroid plexus, the CSF flows to the lateral ventricle, then to the interventricular foramen of Monro, the third ventricle, the cerebral aqueduct of Sylvius, the fourth ventricle, the lateral foramina of Luschka and medial foramen of Magendie, the subarachnoid space, the arachnoid granulations, the dural sinus, and finally into the venous drainage.

COMMUNICATING HYDROCEPHALUS

Communicating hydrocephalus occurs when full communication exists between the ventricles and subarachnoid space.

NON-COMMUNICATING HYDROCEPHALUS (OBSTRUCTIVE HYDROCEPHALUS)

Occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space, resulting in ventricular/subarachnoid space non-communication.

ARRESTED HYDROCEPHALUS

Arrested hydrocephalus is defined as stabilization of known ventricular enlargement, probably secondary to compensatory mechanisms.

CAUSES OF HYDROCEPHALUS

Common Causes of Hydrocephalus in Infants and Children

Congenital Causes

Stenoses of the aqueduct of Sylvius
Dandy-Walker malformation
Arnold-Chiari malformation Type 1 and Type 2
Agenesis of the foramen of Monro
Congenital toxoplasmosis

Acquired Causes

Mass lesions in the posterior fossa
Tumours – medulloblastoma, astrocytoma
Cysts
Abscesses
Haematoma
Intraventricular haemorrhage
Tubercular meningitis
Bacterial meningitis
Cysticercosis
Idiopathic

Causes of Hydrocephalus in Adults

Subarachnoid haemorrhage (SAH)
Idiopathic hydrocephalus
Head injury (through SAH)
Tumours – Ependymoma, choroid plexus papilloma, craniopharyngioma,
Congenital aqueductal stenosis
Meningitis
All causes of hydrocephalus described in infants and children are present in adults who have had congenital or childhood-acquired hydrocephalus.

CLINCIAL FEATURES

History

Symptoms in Infants

Poor feeding
Irritability
Reduced activity
Vomiting

Symptoms in Children

Slowing of mental capacity

Headaches (initially in the morning) that are more significant than in infants because of skull rigidity
Neck pain – suggesting tonsillar herniation
Vomiting – more significant in the morning
Blurring of vision—due to papilloedema
Diplopia – due to sixth nerve palsy
Stunted growth and sexual maturation from third ventricle dilatation: This can lead to obesity and to precocious or delayed onset of puberty.
Difficulty in walking, secondary to spasticity – due to stretching of periventricular pyramidal tract supplying the lower limbs
Drowsiness

Symptoms in Adults

Cognitive deterioration
Headaches
Neck pain
Nausea
Vomiting – more in the morning
Blurring of vision – due to papilloedema
Diplopia due to sixth nerve palsy
Difficulty in walking
Drowsiness
Urinary incontinence

Symptoms of NPH

More Common

Gait disturbances
Dementia
Urinary incontinence

Less Common

Aggressive behavior
Parkinson-like symptoms
Seizures

Physical Examination

Infants

Increase in head circumference
Seperation of sutures – can be seen or palpated clinically
Dilated scalp veins
Tense fontanelles

Setting-sun sign – Both ocular globes are deviated downward, the upper lids are retracted, and the white sclerae may be visible above the iris.
Spasticity preferentially affects the lower limbs

Children

Papilledema
Failure of upward gaze:
Macewen sign or "cracked pot" sound on percussion of the head
Unsteady gait due to spasticity in the lower extremities
Large head with closed sutures due to chronic increase in ICP
Unilateral or bilateral sixth nerve palsy

Adults

Papilledema
Optic atrophy
Failure of upward gaze and accommodation
Unsteady gait is related to truncal and limb ataxia
Spasticity in lower limbs
Unilateral or bilateral sixth nerve palsy is secondary to increased ICP.

NPH

Normal muscle strength
Normal sensory examination
Exaggerated deep tendon reflexes
Positive Babinski response
Difficulty in walking varies from mild imbalance to inability to walk or to stand
Sucking and grasping reflexes appear in late stages.

DIFFERENTIAL DIAGNOSIS

Brainstem gliomas
Craniopharyngioma
Epidural haematoma
Glioblastoma multiforme
Headache
Intracranial haemorrhage
Meningioma
Mental retardation
Pseudotumour cerebri
Subdural empyema
Subdural haematoma
Macrocephaly
Hydranencephaly

Cerebral tumours
Periaqueductal glioma
Agenesis of corpus callosum

RADIOLOGICAL EVALUATION (FIGURE 15.1, 15.2 AND 15.3 A-D)

CT can assess the size of ventricles and other structures.
MRI can evaluate for Chiari malformation or cerebellar or periaqueductal tumours. MRI affords better imaging of the posterior fossa than CT. MRI can differentiate NPH from cerebral atrophy.

MANAGEMENT

Medical Management

Medical treatment is not effective in long-term treatment of chronic hydrocephalus. However treatment with antibiotics is necessary in patients with meningitis.

Surgical Management

Surgical treatment is the preferred therapeutic option. The principle of shunting is to establish a communication between the CSF (ventricular or lumbar) and a drainage cavity (peritoneum, right atrium, pleura). Ventriculoperitoneal (VP) shunt is most commonly perfomed procedure.

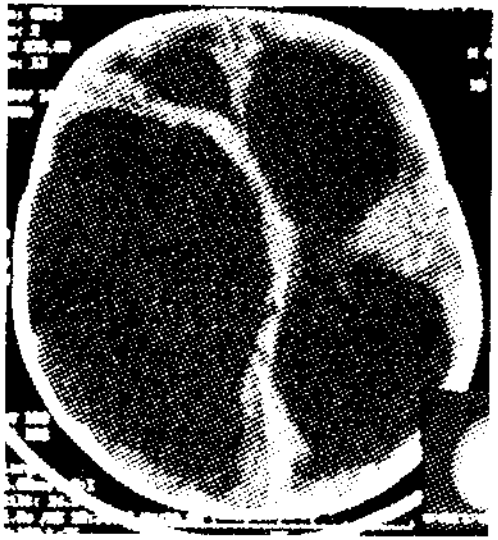


Fig. 15.1 CT scan image of patient with hydrocephalus

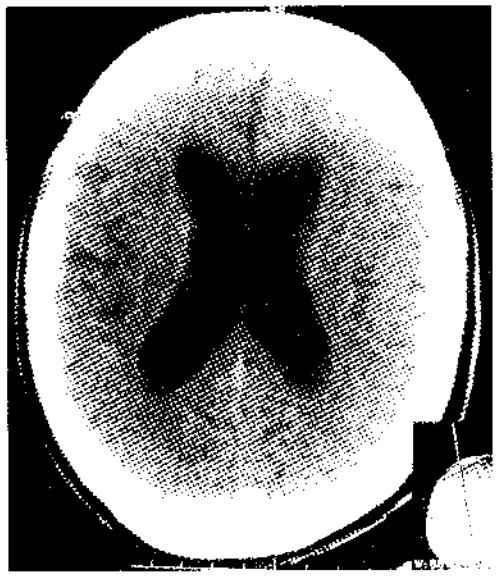
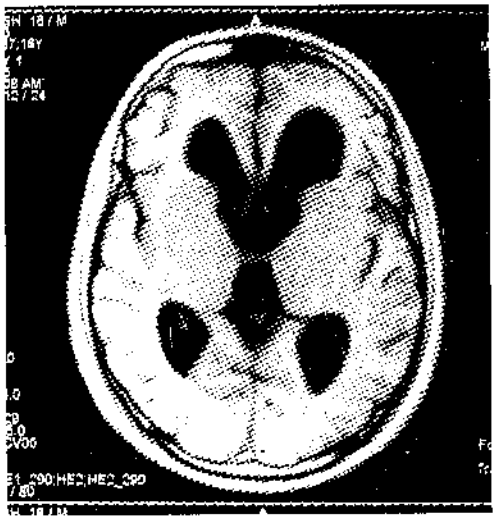


Fig. 15.2 CT scan image of patient with hydrocephalus

A



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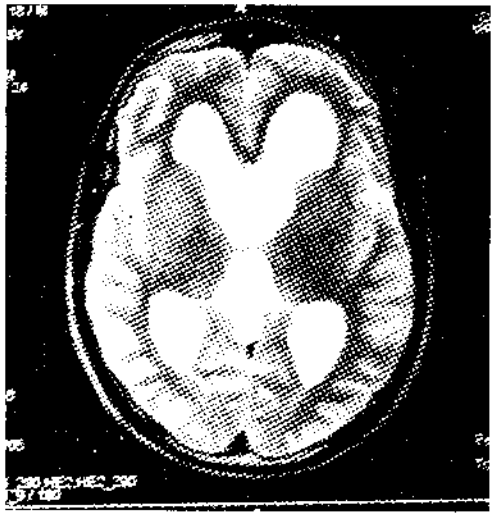

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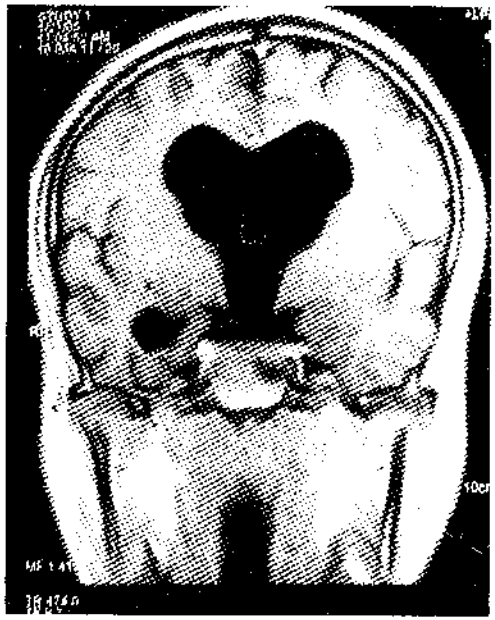

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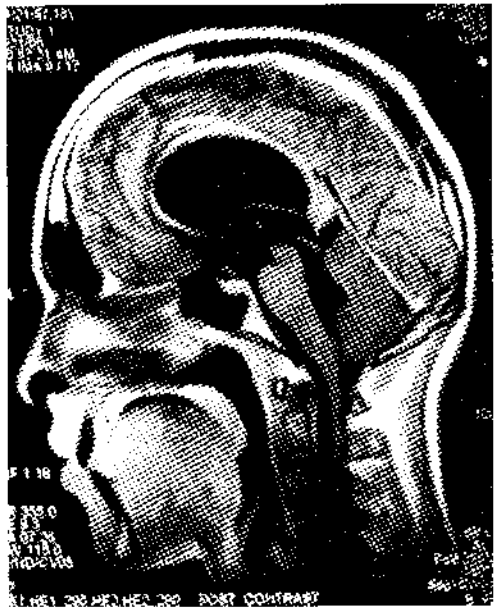


Fig. 15.3 MRI images of patients with hydrocephalus
(A) T1 axial image (B) T2 axial image
(C) T1 coronal image (D) T1 sagittal image

COMPLICATIONS

Related to progression of hydrocephalus
Visual changes
Occlusion of posterior cerebral arteries secondary to downward transtentorial herniation
Chronic papilledema injuring the optic disc
Dilatation of the third ventricle with compression of optic chiasm
Cognitive dysfunction
Incontinence
Gait changes – related to medical treatment
Electrolyte imbalance
Metabolic acidosis
Related to surgical treatment
Shunt infection

PROGNOSIS

Long-term outcome is related directly to the cause of hydrocephalus.

CHAPTER 16

Cerebellar Ataxia

Cerebellar ataxias are a group of disorders due to many different causes. (Box 16.1)

The commonest causes are multiple sclerosis and alcoholic cerebellar disease.

Box 16.1 Causes of cerebellar ataxia

Alcoholism
Multiple sclerosis
Brain tumours
Thyroid disease
Viral infections (chicken pox and Coxsackie during childhood)
Degenerative ataxias
Genetic ataxias
Autosomal dominant cerebellar ataxias
SCA1
SCA2
SCA3 etc
Episodic ataxias
Mitochondrial disorders
NARP (neuropathy, ataxia, and retinitis pigmentosa)
MELAS (mitochondrial encephalomyopathy, lactic acidosis with stroke-like episodes)
MERRF (myoclonus epilepsy with ragged red fibers)
Idiopathic cerebellar ataxia

CLINICAL PRESENTATION

These diseases can manifest mainly in adult life, but also in adolescence or childhood. Certain features differ between patients and may help in making a precise diagnosis.

Common presenting or early manifestations:
- Progressive ataxia of gait (usually broad based)
- Progressive limb ataxia including tremor
- Progressive slurring dysarthria
- Nystagmus

Later on during the course of these diseases:
- Ophthalmoplegia
- Dysphagia
- Parkinsonian features

A minority of patients may encounter:
- Decrease in visual acuity
- Cognitive decline

DIFFERENTIAL DIAGNOSIS

Multiple sclerosis
Posterior fossa tumours
Alcoholic cerebellar ataxia
Ataxia as a non-metastatic manifestation of malignancy
Drug toxicity (e.g., phenytoin)
Vitamin E deficiency

DIAGNOSIS

A careful history may make the diagnosis without further investigation, particularly if there is a clear mode of inheritance within the family.

INVESTIGATIONS

Routine haematology and biochemistry
Liver function tests
Brain imaging
These tests are usually performed to exclude malignancy, vitamin B_{12} deficiency, multiple sclerosis or other causes of ataxia.
Molecular genetic tests may be used to confirm the diagnosis of genetic ataxias.

TREATMENT

Specific Measures

These are aimed at treating the underlying causes (e.g., tumours, alcohol abstinence).

Speech Therapy

Expert speech therapy advice is important, and may lead to alternative communication strategies.

Swallowing

Dysphagia becomes more common as the disease progresses and can lead to weight loss or aspiration. Patients with advanced disease may require palliation with percutaneous endoscopic gastrostomy (PEG).

Depression

All patients with progressive neurological disorders are susceptible to depressive illness. These patients will require counseling and in some cases antidepressants will help to relieve the symptoms.

Genetic Counseling

Genetic counseling is strongly encouraged in families with an incidence of cerebellar ataxia. A thorough genetic and pedigree analysis must be done in order to determine the form of cerebellar ataxia.

Physiotherapy

Physiotherapy is often valuable, particularly to preserve mobility, planning for wheelchair support.

CHAPTER 17

Friedreich's Ataxia

Most common form of inherited ataxia.
Comprising one-half of all hereditary ataxias.
It can occur in its classical form or in association with a genetically determined vitamin E deficiency syndrome.
Inheritance is autosomal recessive.

PATHOLOGY

The primary site of pathology is the spinal cord and the peripheral nerves. There may be slight atrophy of the cerebellum and cerebral gyri. Sclerosis and degeneration are seen predominantly in the spinocerebellar tracts, lateral corticospinal tracts, and posterior columns.

CLINICAL FEATURES

Age of onset—8–16 years
Patients typically presents before the age of 25 years
Common presenting features:
Progressive staggering gait
Frequent falling
Intubation
Lower extremities are more severely involved than the upper extremities
Dysarthria
Rare presenting features
Progressive scoliosis
Foot deformity
Nystagmus
Cardiopathy

NEUROLOGICAL EXAMINATION

Nystagmus
Loss of fast saccadic eye movements
Truncal titubation
Dysarthria, dysmetria, and ataxia of extremity and truncal movements.
Extensor plantar responses
Absent deep tendon reflexes
Weakness (greater distally than proximally)
Loss of vibratory and proprioceptive sensation may occur
Median age of death is 35 years
Women have a significantly better prognosis

ASSOCIATED CONDITIONS

Cardiac involvement occurs in 90 percent of patients with Friedreich's ataxia.
Moderate mental retardation or psychiatric syndromes
High incidence of insulin resistant diabetes (20 percent)
Musculoskeletal deformities(pes cavus, pes equinovarus, and scoliosis)

Section III

DISORDERS OF SPINE AND SPINAL CORD

CHAPTER 18

Spinal Cord Injury

Spinal injuries include fractures, contusions, and compressions of the vertebral column. Eighty per cent of spinal cord injuries occur in males. Approximately 50 per cent are due to road traffic accidents and 25 per cent to domestic and industrial injuries, particularly fall down stairs and from ladders or scaffolding. Sporting accidents, such as those occurring from diving into shallow water, rugby, horse riding, and gymnastics account for at least 15 per cent of injuries.

PATHOLOGY

Spinal cord trauma may result from acceleration, deceleration or other deforming force and can injure the neural tissue by following mechanisms.

Hyperextension

Hyperextension from acceleration-deceleration forces and sudden reduction in the anteroposterior diameter of the spinal cord.

Hyperflexion

Hyperflexion from sudden and excessive force, propelling the neck forward or causing an exaggerated movement to one side.

Vertical

Vertical compression from force being applied from the top of the cranium along the vertical axis through the vertebra.

Rotational

Rotational forces from twisting, which adds shearing forces.

Injury will cause microscopic haemorrhages in the grey matter and pia-arachnoid with subsequent necrosis of neural tissue. Phagocytes appear at the site within 36 to 48 hours after the injury, macrophages engulf degenerating axons, and collagen replaces the normal tissue. Scarring and meningeal thickening leaves the nerves in the area blocked or tangled. In the white matter, circulation usually returns to normal in approximately 24 hours.

CLINICAL FEATURES

Injury to the spinal cord can be classified as complete or incomplete. An incomplete spinal injury may be an anterior cord syndrome, central cord syndrome or Brown-Sequard syndrome, depending on the area of the cord affected (Figure 18.1 A–E). This chart highlights the characteristic signs and symptoms of each (Table 18.1).

Table 18.1 Summary of spinal cord syndromes

Type	Pathology	Clinical features
Complete transection	Complete loss of function below the transaction	Quadriplegia with cervical cord transection Paraplegia with thoracic cord transection Loss of all reflexes and sensory function Bladder and bowel dysfunction Respiratory impairment Loss of vasomotor tone
Central cord syndrome	Center portion of cord affected Due to hype-rextension injury	Weakness greater in upper than in lower extremities Variable degree of bladder dysfunction
Anterior cord syndrome	Occlusion of anterior spinal artery	Loss of motor function below level of injury. Loss of pain and temperature sensations below level of injury. Intact touch, pressure, position and vibration senses
Brown-Sequard syndrome	Hemisection of cord affected due to stabbing and gunshot wounds	Ipsilateral paralysis or paresis below the level of the injury. Ipsilateral loss of touch, pressure, vibration, and position sense below level of injury Contralateral loss of pain and temperature sensations below level of injury

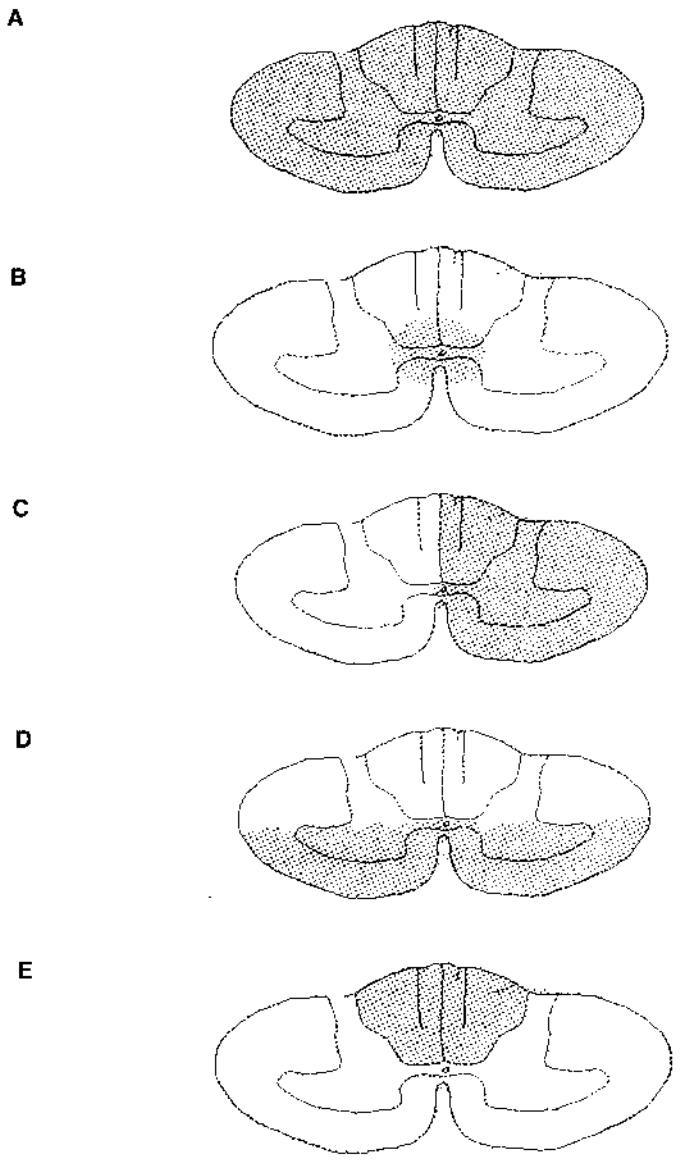


Fig. 18.1
A. Complete transaction
B. Central cord syndrome
C. Brown-Sequard syndrome
D. Anterior cord syndrome
E. Posterior cord syndrome

DIAGNOSIS

Neurological Examination

Detailed neurologic evaluation to locate the level of injury and detects cord damage.

Spinal X-rays

Spinal X-rays (lateral, anteroposterior) will detect the underlying fractures (Figure 18.2 A–D).

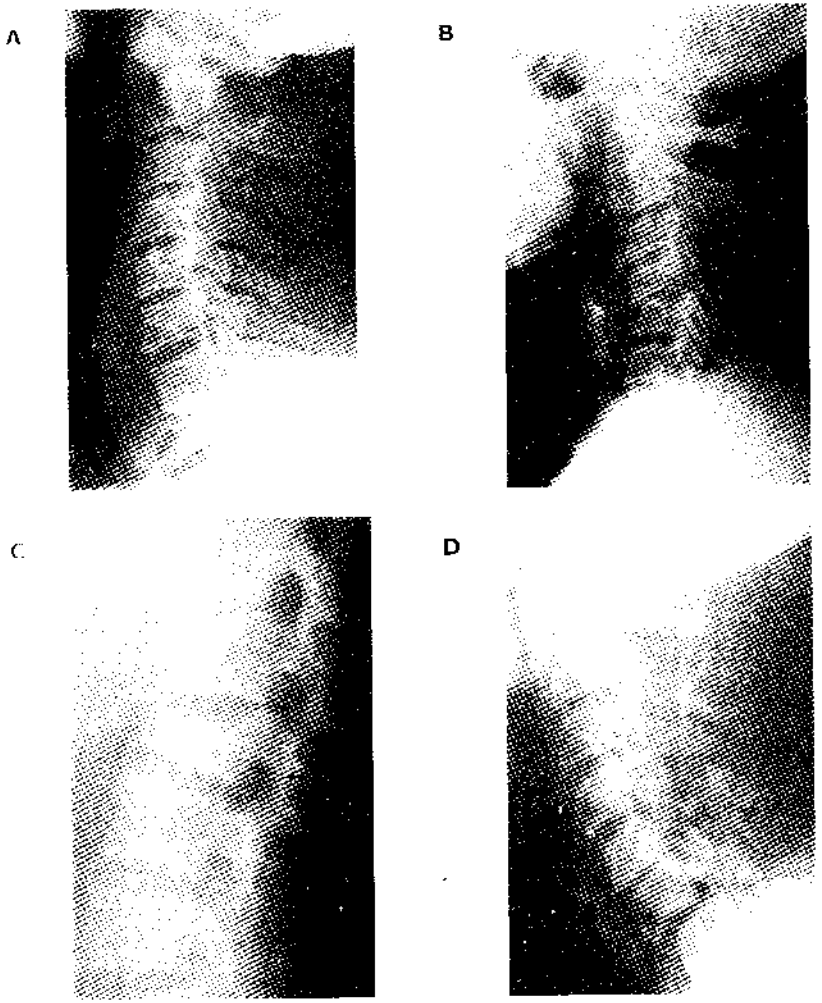
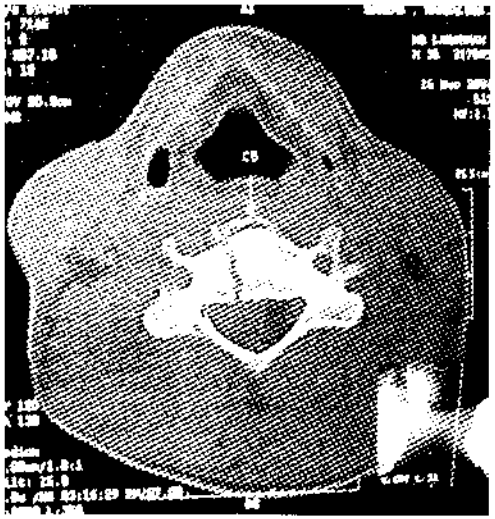


Fig. 18.2 X-rays spine
(A) Atlanto-axial dislocation (B) C5 over C6 anterior subluxation (C) Wedge fracture L1 body (D) Fracture C6 body and C4 5 spinous processes

CT Scan

CT scan will give excellent details of the vertebral column and associated fractures (Figure 18.3 A–C).

A



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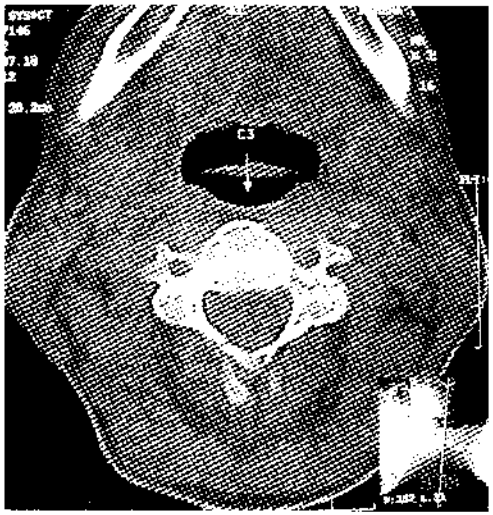


C

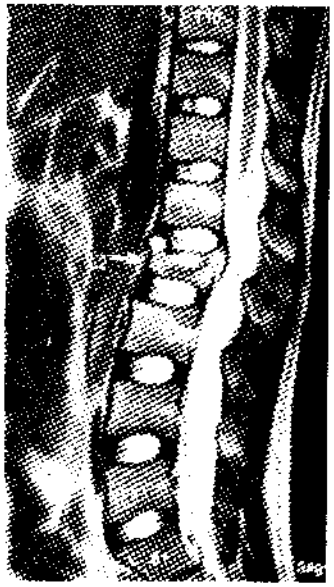
Fig. 18.3 CT scan spine
A. CT scan showing fracture of vertebral body
B. CT scan showing fracture left lamina
C. CT scan showing fracture of odontoid process

Magnetic Resonance Imaging (MRI)

In MRI, bony detail is limited but the spinal cord, and any soft tissue compression will clearly be demonstrated (Figure 18.4 A–D).

A

B



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D

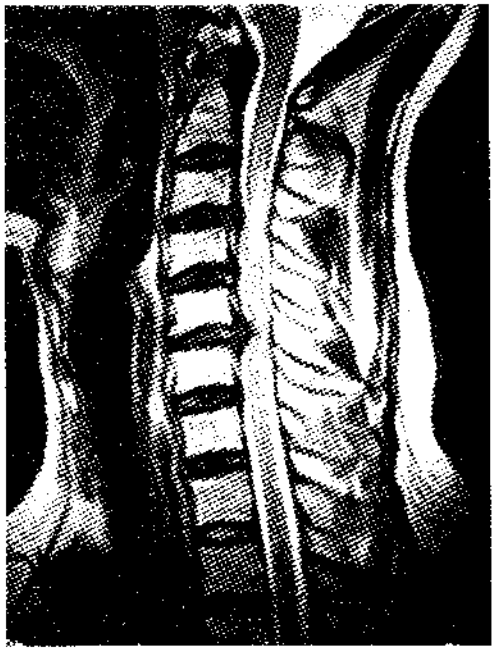


Fig. 18.4 MRI images
(A) C5 over C6 anterior subluxation (B) Fracture of L2 body
(C) MRI showing complete transaction (D) MRI showing cord contusion at C3–4 level and disc prolapse at C5–6 level

MANAGEMENT

The conscious patient with cord damage may complain of neck or back pain, disturbance or loss of sensation, and weakness or flaccid paralysis below a neurological level. In this situation until proven otherwise always suspect spinal cord injury and treat promptly

Immediate Immobilization

Immediate immobilization to stabilize the spine to prevent cord damage (primary treatment). Use of sandbags on both sides of the patient's head, a hard cervical collar, or skeletal traction with skull tongs or a halo device for cervical spine injuries
Maintenance of airway
Intravenous access
Cardiopulmonary support
Treatment of hypoxia and hypotension

Methylprednisolone

High doses of methylprednisolone to reduce inflammation with evidence of cord injury

Bed Rest

Bed rest on firm support (such as a bed board)

Analgesics

To relieve pain inflammation

Urinary Retention

Catheterization (details in Chapter 27)
Bowel care (details in Chapter 27)

Joints and Limbs

The paralysed joints must be passively moved daily, to maintain a full range of movement and prevent contractures, and paralysed hands should be splinted in the position of function.

Pressure Ulcers

The patient should be turned 2-hourly, usually increasing to 3-hourly after a few days if the skin has not marked on 2-hourly turns.

Spasticity (details in Chapter 42)

Management of Associated Injuries

Head Injuries
Chest
Multiple rib fractures
Haemothorax
Pneumothorax
Lung contusion

Limbs Injuries (e.g., fractures)
Reconstructive Surgery Spine
Cervical Spine
Anterior Cervical Arthrodesis (Figure 18.5)
Anterior cervical discectomy with or without interbody fusion
Management of refractory symptoms of cervical disc disease
Low incidence of major complications and postoperative morbidity
High degree of success in relieving symptoms

Techniques
Surgery can be limited to simple discectomy
Surgery can be limited to simple discectomy and interbody fusion
Removal of osteophytes
Decompression of the spinal cord and nerve roots

Fig. 18.5 X-ray cervical spine lateral view showing anterior fixation with screw and plates

Postoperative Complications
Injury to the cervical viscera (trachea, oesophagus)
Neurological and vascular injury
Displacement of a dowel bone graft into the spinal canal
Infection
Retropharyngeal haematoma

Posterior Arthrodesis (Figure 18.6)
Transarticular C1–2 screw fixation

Indications
C1–2 instability
Odontoid fractures

Complications
Screw malpositioning
Vertebral artery injury
Neural injury

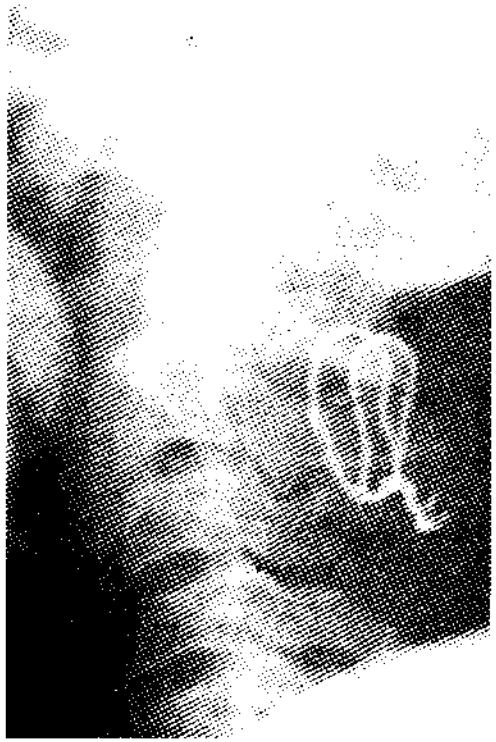


Fig. 18.6 X-ray CV junction showing C1 C2 fixation with sublaminar wiring

Anterior Screw Fixation of Dens Fractures
Internal Fixation and Posterior Plating of Occiput to C2
Triple-Wire Procedure for Posterior Fusion
Anterior Cervical Plating
The development of anterior cervical plates now allows immediate rigid internal fixation after decompression and bone grafting.

Dorsal and Lumbar Spine
Indications
Degenerative, traumatic, and congenital lesions
Posterior, anterior, retroperitoneal, or transperitoneal approach

Posterior arthrodesis (Figure 18.7)
Posterolateral or intertransverse fusions
Internal fixation in spinal fusion
Internal fixation (such as pedicle screws and plates)
Minimally invasive posterior lumbar fusion
Lumbar interbody arthrodesis

Indications for anterior arthrodesis
Debridement of infection, tuberculosis, excision of tumours, correction of kyphosis, scoliosis, neural decompression after fracture, and to achieve stability when posterior arthrodesis is not feasible.
Anterior disc excision and interbody fusion
Minimally invasive anterior fusion

Fig. 18.7 Posterior lumbar fixation with screws and rods

CHAPTER 19

Transverse Myelitis

Transverse myelitis is an acute inflammatory demyelinating disorder affecting the spinal cord. Transverse myelitis is more common in adults than in children.

CLINICAL FEATURES

Transverse myelitis presents with pain at the site of the lesion, followed by weakness in the legs, sensory symptoms, and sphincter involvement. The weakness increases and the clinical picture is that of spinal shock, features which are rarely seen in acute cord lesions due to multiple sclerosis; sphincter control is lost, but unlike patients with multiple sclerosis, there is usually difficulty in emptying rather than filling the bladder.

DIFFERENTIAL DIAGNOSIS

Include structural abnormalities (Tumours and other causes of myelopathy)

INVESTIGATIONS

MRI

MRI of spinal cord may demonstrate cord swelling.

CSF

The spinal fluid shows an increased mononuclear cell count, total protein raised and oligoclonal bands may be present on electrophoresis.
Glucose is usually normal.

TREATMENT

High dose of methylprednisolone

PROGNOSIS

Patients may have near total recovery inspite of severe initial deficits. In some patients it may progress to multiple sclerosis.

CHAPTER 20

Syringomyelia

The essential lesion in syringomyelia is an irregular asymmetrical cavity, filled with cerebrospinal fluid within the spinal cord. The syrinx may extend into the medulla (sryingobulbia).

CAUSES OF SYRINGOMYELIA

Spinal dysraphism (e.g., Arnold-Chiari malformation)
Post-traumatic
Post surgical
Post infectious arachanoid adherences
Spinal cord tumours

PATHOLOGY

The syrinx cavity can extend over many segments, or even its entire length, although usually most extensive in the cervical enlargement. Contiguous structures are destroyed, in particular the anterior horn cells, the sensory fibres decussating within the cord concerned with pain and thermal sensation, and the lateral corticospinal tracts.

CLINICAL FEATURES

The onset of symptoms is usually in early adult life. The first symptoms usually involve one upper limb, with a combination of wasting of the hand muscles, weakness, and sensory loss, often resulting in painless burns, either from cigarettes or from cooking. Sensory loss, classically of dissociated type with preservation of light touch and proprioception, but loss of pain and thermal sensation, often extends over the whole arm and upper thorax or may be bilateral. A Horner's syndrome, or excessive sweating over one side of the face, may indicate involvement of the cervical sympathetic nerve.

Involvement of the brain stem (syringobulbia) will present with sensory loss over the peripheral areas of the face, with sparing of the nose and mouth. Damage to the motor nuclei of the lower cranial nerves causes wasting of the tongue, dysphagia, and vocal cord paralysis. There may be rotatory nystagmus.

INVESTIGATIONS

X-ray Cervical Spine

This may show anomalies around the cranio-vertebral (CV) junction

MRI (Figure 20.1)

The diagnosis of syringomyelia, its extent and causation, can now be fully displayed by magnetic resonance imaging.

A

B

Fig. 20.1 MRI cervical spine showing extensive syringomyelia (a) T1 image, (b) T2 image

TREATMENT

lief of obstruction to the flow of cerebrospinal fluid or the draining of the inx.

CHAPTER 21

Motor Neurone Disease

This group of disorders is characterized clinically by weakness and variable wasting of affected muscles, without accompanying sensory changes. Motor neurone disease in adults generally commences between 30 and 60 years of age. The motor neurone diseases result from selective loss of function of the lower and/or upper motor neurons controlling the voluntary muscles of the limbs or bulbar region. Sensation and cognition are normal on simple clinical assessment in the motor neurone diseases.

CLASSIFICATION

Five varieties have been distinguished on clinical grounds.

Progressive Bulbar Palsy

Bulbar involvement predominates owing to disease processes affecting primarily the motor nuclei of the cranial nerves.

Pseudobulbar Palsy

Bulbar involvement predominates in this variety also, but it is due to bilateral corticobulbar disease and thus reflects upper motor neurone dysfunction.

Progressive Spinal Muscular Atrophy

This is characterized primarily by a lower motor neurone deficit in the limbs due to degeneration of the anterior horn cells in the spinal cord.

Primary Lateral Sclerosis

There is a purely upper motor neuron deficit in the limbs.

Amyotrophic Lateral Sclerosis

A mixed upper and lower motor neurone deficit is found in the limbs. This disorder is sometimes associated with dementia or parkinsonism.

CLINICAL FEATURES

At presentation, patients either have bulbar or spinal symptoms, although both usually become evident as the disease progresses.

The Bulbar Form

Difficulty in swallowing, chewing, coughing, breathing, and talking (dysarthria) occur with bulbar involvement. In progressive bulbar palsy, there is drooping of the palate, a depressed gag reflex, pooling of saliva in the pharynx, a weak cough, and a wasted, fasciculating tongue.

In pseudobulbar palsy, the tongue is contracted and spastic and cannot be moved rapidly from side to side. Limb involvement is characterized by motor disturbances (weakness, stiffness, wasting, fasciculations) reflecting lower or upper motor neurone dysfunction; there are no objective changes on sensory examination, though there may be vague sensory complaints. The sphincters are generally spared.

The Spinal Form

The spinal form of amyotrophic lateral sclerosis usually presents with wasting and weakness of one limb, usually as intrinsic hand muscle wasting or foot drop. Asymptomatic involvement of other limbs is often evident on examination. It is diagnostically important to demonstrate combined upper and lower motor neurone signs in at least two limbs. Wasted fasciculating muscles also exhibiting clonus or hyper-reflexia are a helpful finding.

INVESTIGATIONS

Electromyography

Electromyography may show changes of chronic partial denervation, with abnormal spontaneous activity in the resting muscle and a reduction in the number of motor units under voluntary control.

Nerve Conduction Studies

Motor conduction velocity is usually normal but may be slightly reduced, and sensory conduction studies are also normal.

Biopsy

Biopsy of a wasted muscle shows the histological changes of denervation.

Serum Creatine Kinase

The serum creatine kinase may be slightly elevated but never reaches the extremely high values seen in some of the muscular dystrophies.

CSF

The cerebrospinal fluid is normal.

DIFFERENTIAL DIAGNOSIS

Spondylitic radiculomyelopathy
Postpolio syndrome
Neurolathyrism

TREATMENT

No treatment is known to cure amyotrophic lateral sclerosis. Yet much can be done to overcome disability and alleviate distress by the care team of speech therapist, physiotherapist, occupational therapist, social worker, and physician.

Drugs

Riluzole

This drug reduces the presynaptic release of glutamate and may slow the progression of disease.

Supportive Treatment

Anticholinergic drugs (such as trihexyphenidyl, amitriptyline, or atropine)- For troublesome drooling
Braces or a walker to improve mobility
Physical therapy to prevent contractures
Spasticity (discussed in chapter)
Dysphagia – semiliquid diet or nasogastric tube
In extreme cases gastrostomy or cricopharyngomyotomy is necessary to support nutrition.
Tracheostomy may be necessary if respiratory muscles are severely affected.

PROGNOSIS

The disorder is progressive, and amyotrophic lateral sclerosis is usually fatal within 3–5 years; death usually results from pulmonary infections. Patients with bulbar involvement generally have the poorest prognosis.

CHAPTER 22

Spina Bifida

The nervous system develops by the formation of a tubular structure (neurulation), and closure of this tube is completed by closure of the cranial and caudal neuropores at about days 24 to 26 of gestation. Failure of fusion or rupture of a previously closed neural tube can result in different type of congenital anomalies (Box 22.1).

Box 22.1

Meningocele
Myelomeningocele
Lipomyelomeningocele
Low lying conus
Diastematomyelia Thickened filum terminale

ASSOCIATED CONDITIONS

Hydrocephalus – 80% to 90% have hydrocephalus that requires cerebrospinal shunting
Hydrosyringomyelia
Arnold-Chiari malformation Type I and Type II
Myelomeningocele is the most common of the spectrum of conditions (Table 22.1). A myelomeningocele is a sac-like structure containing cerebrospinal fluid and neural tissue. The hernial protrusion of the spinal cord and its meninges through a defect in the vertebral canal results in variable neurological defects depending on the location and severity of the lesion.

Table 22.1 Classification of myelomeningocele

Group	Level of lesion	Resultant muscle function
Group I	Thoracic or high lumbar	No quadriceps function Community ambulation is rare
Group II	Low lumbar	Functioning quadriceps and medial hamstring muscles No gluteus medius function Most children in this group require ankle-foot orthoses for support and crutches for trunk stability
Group III	Sacral	Functioning quadriceps and gluteus medius muscles Most children in this group can walk without external support and may or may not require ankle-foot orthoses

TETHERED CORD SYNDROME

Clinical signs are variable and depend on the site of lesion and associated anomalies. These include:
Loss of motor function
Development of spasticity in the lower extremities, primarily the medial hamstrings and ankle dorsiflexors and evertors
Development of scoliosis before the age of 6 years in the absence of congenital anomalies of the vertebral bodies
Back pain and increased lumbar lordosis in an older child
Bowel and bladder dysfunction

INVESTIGATIONS

X-ray

X-ray may show the bony anomalies and scoliosis (Figure 22.1)

MRI (Figure 22.2)

Magnetic resonance imaging (MRI) shows signs of tethering of the spinal cord in most children with myelomeningocele.

Computed Tomography (CT) (Figure 22.3)

Myelographic evaluation
Prenatal diagnosis can be made by biochemical and enzyme evaluation, as well as by roentgenographic or ultrasound examination.

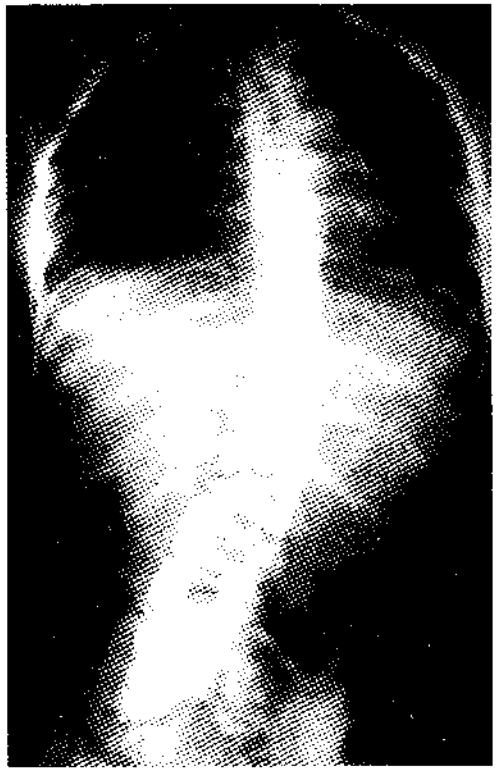


Fig. 22.1 X-ray dorso-lumbar spine showing scoliosis

Fig. 22.2 MRI lumbar spine showing defect posteriorly in spina bifida patient

A

B

Fig. 22.3 CT scan of the same patient showing
(A) Bifid lamina, (B) Bony septum dividing the canal

CLINICAL EVALUATION OF CHILDREN WITH MYELOMENINGOCELE

Serial sensory and motor examinations to evaluate the neurological level of function.

Sitting balance as an indication of central nervous system function. If one or two hands are required for support while sitting, the probability of ambulation is significantly decreased.

Upper extremity function, including decreased grip strength and atrophy of the thenar musculature (suspect hydromyelia).

Presence or absence of scoliosis, kyphosis, and increased lumbar lordosis.
Range of motion, stability, and contractures of the hip.

Alignment, range of motion, contractures, and spasticity of the knee.
Rotational deformities, including external tibial torsion.

Ankle valgus deformity.

Other foot deformities, including congenital vertical talus or varus foot deformity.

GOALS OF MANAGEMENT

Stable posture
Ambulation (may not be possible for every child)
Effective ambulation with low energy consumption and minimal bracing
Activities of daily living without lower extremity function
Functional independence

Almost all patients with L2, or higher-level lesions are wheelchair users, and more than two-thirds of those with lower level lesions (L3 to L5) use a wheelchair at least part of the time.

Prerequisites for Walking

A spine balanced over the pelvis
Absence of hip and knee contractures (or only mild contractures)
Plantigrade, supple, braceable feet with the center of gravity centered over them.

Orhtotic Management

Aim of orthotic treatment is to obtaining effective mobility with minimal restriction.

Bracing and splinting vary with the degree of motor deficit and trunk balance, and each child should be carefully evaluated by the orthopaedic surgeon, the orthotist, and the physical therapist. Children aged 12 to 18 months may benefit from the use of an A-frame for standing, and for children over 2 years of age, a parapodium supports the spine and allows a swing-to or swing-through gait with crutches or a walker. An ankle-foot orthosis (AFO) is used in children with low lumbar or sacral level lesions and fair quadriceps muscle function. The AFO should be rigid enough to provide ankle and foot stabilization and to maintain the ankle at 90 degrees. A knee-ankle-foot orthosis (KAFO) may be indicated for a child with

a lumbar level lesion and weak quadriceps function to prevent abnormal valgus of the knee during the stance phase of gait. Children with high-level lesions often have excessive anterior pelvic tilt and lumbar lordosis and require a pelvic band, either as a conventional hip-knee-ankle-foot orthosis (HKAFO) or a reciprocating gait orthosis. The reciprocating gait orthosis also can be used in patients with upper lumbar lesions, allowing them to be upright and assisting them in attempts at ambulation. This brace provides the ability to walk in a reciprocal fashion by dynamically coupling the flexion of one hip to the simultaneous extension of the contralateral hip. For the reciprocating orthosis to be effective, the patient should have good upper extremity strength, trunk balance, and active hip flexion.

Surgical Management

If clinical signs are documented, surgical treatment is indicated to prevent further deterioration of the motor function and to diminish the progress of spasticity and scoliosis.

It is important to make an early diagnosis and start treatment because surgical release of the tethered cord rarely provides complete return of lost function.

Procedures

Contracture release
Tendon transfer
Spasticity
Arthrodesis

CHAPTER 23

Degenerative Disc Diseases

PROLAPSED INTERVERTEBRAL DISC DISEASE

A disc prolapse consists of an acute or subacute backwards dislocation of disc substance.

Pathology

Disc protrusions often follow an episode of sudden or unusual exertion leading to tear of posterior longitudinal ligament. Protrusion of disc material into the spinal canal can cause nerve root or cord compression.

Clinical Features of Nerve Root Lesions

Irritation or compression of a root may give rise to pain characteristically made worse by movement of the spine and by actions which cause sudden pulses of pressure in the spinal subarachnoid space, such as coughing or sneezing.

Pain and paraesthesiae in the sensory distribution of the involved root
Motor, sensory, and reflex loss.

Cervical Disc Protrusions

Commonest at the C4/5, C5/6, and C6/7 levels, compressing the C5, C6, and C7 roots, respectively.
Pain and stiffness in the neck
Painful limitation of some neck movements

Differential Diagnosis

Median and ulnar nerve palsies
Lesions of the rotator cuff of the shoulder joint

Involvement of the lower part of the brachial plexus by an apical carcinoma of the lung (Pancoast's syndrome)

Dorsal Disc Protrusions

Extremely rare

Lumbar Disc Protrusions

Commonest disc protrusions are at the two lowest levels, L4/5 and L5/S1.
Acute low back pain and sciatica
An L4/5 disc protrusion compresses the L5 root and L5/S1 disc protrusion compresses the S1 root.
The neurological changes correspond to the root affected and vary according to how severely it is compressed.
L5 root involvement leads to weakness of dorsiflexion and eversion of the ankle, numbness on the dorsum of the foot and lateral shin, but no reflex loss.
S1 root involvement causes numbness of the outer border of the foot and little toe, weakness of plantar flexion at the ankle and a reduced or absent ankle jerk.

Plain Radiographs

Plain radiographs are of little value in predicting the level of a symptomatic protrusion.
It will show degenerative changes (Figure 23.1)
May show bone destruction due to tumour or inflammatory lesions

MRI (Figure 23.2 and 23.3)

Investigation of choice as it will give the maximum information.

CT Myelography

CT myelography is especially useful for patients who have already undergone spinal surgery or those where the pathological changes are not especially marked.

Treatment

Cervical Disc Protrusion

Cervical collar
In majority of the cases symptoms abate within a few weeks with conservative measures
Rest
Analgesics
Surgery is reserved for the patients with an incapacitating degree of pain, severe root compression with disabling motor or sensory loss.

Fig. 23.1 X-ray cervical spine showing anterior osteophytes at C4, C5 and C6 level with degenerative changes

Fig. 23.2 MRI cervical spine showing disc prolapse at C5–6 level with compression of thecal sac

Fig. 23.3 MRI lumbo-sacral spine showing disc prolapse at L4–5 level with compression of thecal sac

Lumbar Disc Protrusions

Bed rest for 2–3 weeks

Surgery is only indicated if there is prolonged or recurrent incapacitating root compression or in those cases where serious motor weaknes.

Absolute indication for surgery is a central protrusion compressing the cauda equina.

CHAPTER 24

Spinal Tumours

Spinal cord neoplasms are uncommon lesions affecting only a minority of the population. However, with growth, these lesions can cause limb dysfunction, motor and sensation loss, and possibly, death. These neoplasm can affect vertebral column or may arise from spinal cord and its coverings (Table 24.1).

Table 24.1 Classification of brain tumours

Vertebral column	Metastases Thyroid Breast Lung GI tract Renal Prostate
	Primay bone tumours ***Benign*** Haemangioma Osteoid osteoma Osteoblastoma Aneurysmal bone cyst Giant cell tumour ***Malignant*** Myeloma Lymphoma
Spinal cord and coverings	Ependymomas Astrocytomas Meningioma Schwannomma Haemangioblastoma Metastases

CLINICAL FEATURES

Pain

Usually localised to the anatomical location of the tumour, the pain is progressive over weeks or months, although it may fluctuate in intensity.

Local Tenderness

Local tenderness may be present but this is more common with extraspinal tumours.

Radicular Signs

Owing to the root irritation this may result in pain in a radicular distribution with associated pins and needles in the corresponding dermatomal area. Clinical examination may reveal signs of a radiculopathy — indicated by a lower motor neuron lesion of the affected nerves.

Spinal Cord/Cauda Equina Signs

Clinical signs and symptoms are dependent upon the level of the tumour.

Above the L1/L2 Junction
Progressive spastic paraparesis/quadriparesis
Ascending sensory level
Signs of an upper motor neuron lesion
Perianal and pericoccygeal sensation may be spared

Cauda Equina Compression (below L1/L2 junction)
Lower motor neuron lesion with radicular pain
Dermatomal sensory loss
Myotomal loss of power
Reflexes will be reduced
Normal or absent plantar responses

INVESTIGATIONS

Blood Tests

Complete blood cell count
Prothrombin time/activated partial thromboplastin time

Plain Radiographs

Abnormal findings in 20% of patients
Vertebral collapse
Loss of bone substance
Scalloping of the vertebral bodies on lateral radiographs

Widening of interpedicular distance on anteroposterior radiographs
Scoliosis in children resulting from neuromuscular impairment

MRI (Table 24.2)

MRI with and without gadolinium enhancement is the investigation of choice for spinal tumours. It is the most sensitive and reliable way of diagnosing spinal tumours, and allows assessment of the size and invasiveness of the tumour. Figure 24.1 A,B, Figure 24.2, and Figure 24.3 A,B.

A

B

Fig. 24.1 MRI showing spinal tumour involving cervico-medullary region (A) T1 image, (B) T2 image

Fig. 24.2 MRI showing spinal tumour at C2, 3–4 level (dorsally located)

A

B

Fig. 24.3 MRI showing tumour involving vertebral body with compression of thecal sac (A) T1 image, (B) T2 image

Table 24.2 MRI features of common spinal tumours

Tumour type	T1 W images	T2 Waimages	Contrast homogenous	Cyst formation
Ependymoma	Isointense signal with spinal cord	Hyperintense signal	Strong homogeneous enhancement	
Astrocytoma	Isointense or hypointense signal with spinal cord	Hyperintense signal	Heterogeneous enhancement	Present
Hemangiobl-astoma	Isointense signal to spinal cord	Hyperintense signal	Enhances strongly	Cystic with tumour nodule

CT Scan

CT scans can also be used for better definition of bone and bone destruction.
Nonspecific spinal canal and spinal cord widening
Scalloping of vertebral bodies (intraparenchymal syringomyelia)

Myelogram

Nonspecific spinal canal and spinal cord widening
Multisegmental involvement
Block of contrast dye
Conus region lesions, possible meniscus around the tumour

Spinal Arteriogram

This may be beneficial only if a hemangioblastoma is suggested as a differential diagnosis. Hemangioblastoma arteriogram findings include a vascular blush with a prominent draining vein.

Bone Scans

May show turnouts, although they are unreliable in myeloma and plasmacytoma

Baseline Urodynamics

Findings may assist in diagnosing abnormal bladder function.

TREATMENT

Treatment of spinal tumour is aimed at:
Relief of pain
To achieve a histological diagnosis
Excision of benign tumours
Prevention of further neurological deterioration.

CHAPTER 25

Poliomyelitis

Poliomyelitis is a highly contagious infectious disease caused by three types of poliovirus (P1, P2, and P3).

PATHOGENESIS

The virus enters through the mouth and primary multiplication of the virus occurs at the site of implantation in the pharynx and gastrointestinal tract. The virus is usually present in the throat and in the stool before the onset of illness. The virus invades local lymphoid tissue, enters the blood stream, and then may infect cells of the central nervous system. Replication of poliovirus in motor neurons of the anterior horn and brain stem results in cell destruction and causes the typical manifestations of poliomyelitis. The incubation period for poliomyelitis is commonly 6 to 20 days with a range from 3 to 35 days.

CLINICAL TYPES

Inapparent Infection

The majority of individuals (90–95 percent) have no symptoms at all. This is referred to as inapparent infection.

Abortive Poliomyelitis

A mild and short course of the disease with one or more of the following symptoms:
Fever (up to 103°F or 39.5°C)
Decreased appetite
Nausea and/or vomiting
Sore throat
Not feeling well all over

onstipation
bdominal pain

Nonparalytic Poliomyelitis

The symptoms for nonparalytic poliomyelitis are the same as abortive oliomyelitis but the headache, nausea, and vomiting may be worse. In ddition the following symptoms may occur:
ain of the muscles in the neck, trunk, arms, and legs
tiffness in the neck and along the spine.

Paralytic Poliomyelitis

The symptoms for paralytic poliomyelitis are the same as nonparalytic and bortive poliomyelitis. In addition, the following symptoms may occur:
Muscle weakness all over
Severe constipation
Muscle wasting
Weakened breathing
Difficulty swallowing
Weak cough
Flushed or blotchy skin
Hoarse voice
Bladder paralysis
Muscle paralysis

INVESTIGATIONS

Viral Isolation

Poliovirus may be recovered from the stool or pharynx of a person with poliomyelitis. Isolation of virus from the cerebrospinal fluid (CSF) is diagnostic, but is rarely accomplished.

Serology

Neutralizing antibodies appear early and may be at high levels by the time the patient is hospitalized and, therefore, a 4-fold rise may not be demonstrated.

Cerebrospinal Fluid (CSF)

Increased white blood cells (10 to 200 cells/mm^3, primarily lymphocytes) Mildly elevated protein from 40 to 50 mg/100 mL.

TREATMENT

While there is prevention of the poliomyelitis, there is no cure for individuals who become infected. Treatment is mostly supportive.

Analgesics
To reduce pain and for control of temperature

Bed Rest
Minimal exertion and exercise

PREVENTION

Immunization (IPV – Inactivated polio vaccine or OPV – Oral polio vaccine is preferred)

PHYSIOTHERAPY AND ORTHOPAEDIC MEASURES FOR COMPLICATIONS (SEE CHAPTER 45 & 48)

CHAPTER 26

Subacute Combined Degeneration of the Spinal Cord

This is the commonest and most serious neurological complication of the deficiency of vitamin B_{12}.

PATHOLOGY

As the name suggests there is combined degeneration observed at autopsy in the lateral and posterior columns of the spinal cord on histopathology.

CLINICAL FEATURES

Age range – predominantly middle aged or older.
Symmetrical paraesthesiae in the extremities, usually initially in the feet but occasionally confined to the hands.
Loss of postural sense causes ataxia in walking
Examination in the early stages show distal cutaneous sensory loss
Severe loss of postural sense in the lower limbs
Absent ankle jerks

Extensor platar reflexes
If the disease is left untreated, the disease will progress to severe paraplegia, fatal within a year.

Other Neurological Findings

Moderate loss of mental acuity
Visual impairment

DIAGNOSIS

Clinical evidence of anaemia

Peripheral blood smear may show megaloblastic anaemia
Bone marrow also may show megaloblastic hyperplasia
Serum vitamin B_{12} – almost always significantly reduced

DIFFERENTIAL DIAGNOSIS

Peripheral neuropathy from other causes
Multiple sclerosis
Tingling in the hands alone may be mistaken for the carpal tunnel syndrome.

TREATMENT

Hydroxocobalamin, 1 mg, should be injected immediately, folllowed by every week for several weeks continued indefinitely to avoid relapse.

PROGNOSIS

The response to treatment is in general, excellent.
Even bedridden patients can be restored to normal walking, and extensor plantar reflexes revert to flexor.
Beyond a certain point of disability, however, treatment is unavailing, presumably because of axonal degeneration.

CHAPTER 27

Disorders of Autonomic Nervous System

Disorders of the ANS may result from central nervous system (CNS) or peripheral nervous system (PNS). Causes; they may be generalized, segmental, or focal.

CLASSIFICATION

Generalized ANS Disorders

Multiple system atrophy (Shy-Drager syndrome, Olivopontocerebellar degeneration)
Parkinson's disease
Huntington's disease
Hypothalamic disorders
Pure autonomic failure
Guilliane-Barre syndrome
Raynaud's syndrome

Peripheral ANS Disorders

Peripheral neuropathy (diabetes, amyloidosis, porphyria)
Spinal cord and root lesions
Guilliane-Barre syndrome
Tabes dorsalis

Focal ANS Disorders

Horner's syndrome
Causalgia

CLINICAL FEATURES

Clinical signs and symptoms are due to interruption of the reflex arc controlling autonomic responses. The clinical manifestations of autonomic lesions are influenced by the organ involved, the normal balance of sympathetic-parasympathetic innervation, the nature of the underlying illness, and the severity and stage of progression.

Summary of Clinical Features in Autonomic Dysfunctions and their Management

- Orthostatic hypotension
  - Small meals
  - Avoid alcohol intake and excessive environmental temperatures
  - Increase salt intake
  - Sleeping in a reverse Trendelenburg position (head-up tilt)
  - Compressive garments are of questionable value
  - Drug therapy – Fludrocortisone
  - Other drugs – beta-blockers (propranolol, pindolol, xamoterol), sympathomimetics (ephedrine, midodrine), dopamine antagonists (metoclopramide), and venoconstrictors (dihydroergotamine)
- Postural dizziness – elastic stockings, ephedrine, tyrosine, beta-blockers, mineralocorticoids e.g., fludrocortisone 0.1–0.3 mg daily.
- Reflux oesophagitis and delayed gastric emptying – metoclopramide 10 mg before meals.
- Nocturnal diarrhoea – metoclopramide 10 mg, 8 hourly, a short course of tetracycline may be of benefit to the patient.
- Post gustatory sweating – anticholinergics – propantheline hydrobromide before meals.
- Bladder dysfunction and urinary retention – regular voiding and antibiotics for infections.
- Impotence – counseling, pharmacological erections and penile implants.
- Cardiorespiratory arrest – maintain high FiO_2 at all times.
- Pupillary abnormalities
- Thermal irregularities
- Skin colour abnormalities.

DIAGNOSIS

Diagnosis of the site of reflex interruption is dependent on associated clinical findings, autonomic nervous system tests, and neuroimaging.

Autonomic functions testing

Heart Rate Variation with Deep Breathing

This is a test of parasympathetic influence on cardiovascular function.

Valsalva Response

This response assesses integrity of the afferent limb, central processing, and efferent limb of the baroreceptor reflex.

Heart rate variation during quite inspiration and expiration is lost with autonomic dysfunction.

Postural Hypotension

On standing, the decreased blood pressure is usually compensated for by reflex tachycardia and vasoconstriction, neither of these mechanisms work well in autonomic dysfunction leading to a marked difference in **systolic** blood pressure between lying and sitting/standing. The normal is 10 mm Hg, pathological is when the difference is > 30 mm Hg.

Orthostatic Blood Pressure Recordings

Beat-to-beat BP measurements determined in supine, 80° tilt, and tilt-back positions are useful to quantitate orthostatic failure in BP control. It important to allow a 20-min period of supine rest before assessing changes BP during tilting.

Blood Pressure Response to Sustained Handgrip

Sustained handgrip as measured by a dynamometer causes a reflex increase heart rate and cardiac output without changing systemic vascular resistance, **diastolic** blood pressure thus normally increases.

Sudomotor Function

The capacity to produce sweat can be assessed quantitatively or qualitatively. A reduced or absent response indicates a lesion of the postganglionic sudomotor axon.

Cold Pressor Test

The cold pressor test assesses sympathetic function by having the subject immerse one hand in ice water (1 to 4°C) and then measuring BP at 30s and 1 min. The systolic and diastolic pressures normally rise by 10 to 20 mm Hg. The afferent pathway is spinothalamic and thus is distinct from the afferent limb of the baroreceptor reflex arc. When spinothalamic pathways are intact, an abnormal response indicates an abnormality of autonomic central processing or sympathetic efferent function.

TREATMENT

Management of autonomic failure is limited to alleviating the disability caused by the symptoms. If possible, the primary disorder should be treated, that may not improve autonomic functions.

BLADDER DYSFUNCTIONS

The anatomy and physiology of the bladder are arranged to allow storage of urine until it can be conveniently excreted. Optimal function requires a balance between the bladder and the urogenital musculature. In neurologic disease, the balance among urethral pressure at the bladder neck, external sphincter and pelvic muscle tension, and detrusor pressure is disturbed, which can cause incomplete or unexpected voiding.

Normal Bladder Function

Anatomical Considerations

Sensory Innervation

The majority of the detrusor muscle derives its innervation via parasympathetic fibers (S2–S4). The area of the trigone, however, is innervated by sympathetic fibers (T11–L2). Both somatic sensory fibers and parasympathetic ascending fibers leave the bladder to converge at the S2–S4 levels. There are also some ascending sympathetic fibers that synapse at the T9–L2 levels. The lateral spinothalamic tracts and the fasciculus gracilis carry the ascending fibers to higher centers.

Motor Innervation

The corticospinal tracts carry motor fibers to the external sphincter and pelvic floor muscles, which are under voluntary control. The primitive micturition reflex is at the S2–S4 level via parasympathetic efferent fibers.

Reflex Control

Bladder Smooth Muscles

The bladder smooth muscle has some inherent contractile activity; however, when its nerve supply is intact, stretch receptors in the bladder wall initiate a reflex contraction that has a lower threshold than the inherent contractile response of the muscle.

Sensory Innervation

Fibers in the pelvic nerves are the afferent limb of the voiding reflex, and the parasympathetic fibers to the bladder that constitute the efferent limb also travel in these nerves. The reflex is integrated in the sacral portion of the spinal cord. In the adult, the volume of urine in the bladder that normally initiates a reflex contraction is about 300–400 mL. The sympathetic nerves to the bladder play no part in micturition, but they do mediate the contraction of the bladder muscle that prevents semen from entering the bladder during ejaculation.

Motor Control

There is a facilitatory area in the pontine region and an inhibitory area in the midbrain. After transection of the brain stem just above the pons, the threshold is lowered and less bladder filling is required to trigger it, whereas after transection at the top of the midbrain, the threshold for the reflex is essentially normal. There is another facilitatory area in the posterior hypothalamus. In humans with lesions in the superior frontal gyrus, the desire to urinate is reduced and there is also difficulty in stopping micturition once it has commenced.

Physiology of Micturition

The smooth muscles of the bladder are arranged in spiral, longitudinal, and circular bundles. Contraction of this muscle, which is called the detrusor muscle, is mainly responsible for emptying the bladder during urination (micturition). Micturition is fundamentally a spinal reflex facilitated and inhibited by higher brain centers and, like defecation, subject to voluntary facilitation and inhibition. Urine enters the bladder without producing much increase in intravesical pressure until the viscus is well filled. The first urge to void is felt at a bladder volume of about 150 mL, and a marked sense of fullness at about 400 mL.

During micturition, the perineal muscles and external urethral sphincter are relaxed; the detrusor muscle contracts; and urine passes out through the urethra. The perineal muscles and external sphincter can be contracted voluntarily, preventing urine from passing down the urethra or interrupting the flow once urination has begun. After urination, the female urethra empties by gravity. Urine remaining in the urethra of the male is expelled by several contractions of the bulbocavernosus muscle.

Neurological Causes of Bladder Dysfunction

Suprapontine Lesions

Suprapontine lesions lead to uninhibited bladder contractions possibly secondary to loss of cerebral cortex inhibition at the sacral micturition center. Facilitation of the spinobulbospinal reflex also is affected.

Cerebrovascular accidents
Multiple sclerosis
Dementia
Brain tumours
Traumatic brain injury

Suprasacral Lesions

These lesions cause interruption of the spinobulbospinal reflex, which leads acutely to areflexia, then usually to detrusor hyper-reflexia and uncoordinated micturition with detrusor sphincter dyssynergia.

Trauma
Tumours
Spina bifida

Sacral Lesions (conus medullaris, cauda equine, S2–S4 peripheral nerves)

Lesions of this type lead to variable loss of parasympathetic and somatic nerve function. Detrusor areflexia, bladder neck incompetence, and/or loss of external sphincter function may occur.

Trauma
Stenosis
Tumours
Peripheral neuropathy

Other Causes

Urinary outflow obstruction
Prostatism
Urinary tract infection

Abnormalities of Micturition (Table 27.1)

There are three major types of bladder dysfunction due to neural lesions

1) The type due to interruption of the afferent nerves from the bladder
2) The type due to interruption of both afferent and efferent nerves
3) The type due to interruption of facilitatory and inhibitory pathways descending from the brain.

In all three types the bladder contracts, but the contractions are generally not sufficient to empty the viscus completely, and residual urine is left in the bladder.

Clinical Examination

Details of neurological event has led to symptomatology (e.g., spinal cord injury, multiple sclerosis etc.)
Establish premorbid voiding function and symptoms

Table 27.1 Summary of effects of different injuries on bladder function

Type of injury	Effects on bladder function
Deafferentation	Injury to sacral dorsal roots All reflex contractions of the bladder are abolished The bladder becomes distended, thin-walled, and hypotonic
Denervation	When the afferent and efferent nerves are both destroyed Tumours of the cauda equina or filum terminale The bladder is flaccid and distended
Spinal cord transection	***During spinal shock*** The bladder is flaccid and unresponsive Becomes overfilled, and urine dribbles through the sphincters (overflow incontinence) ***After spinal shock*** The voiding reflex returns No voluntary control and no inhibition or facilitation from higher centers when the spinal cord is transected Bladder capacity is reduced, and the wall becomes hypertrophied Also called the spastic neurogenic bladder

Physical Examination

Determine the motor level of the lesion
Determine sensory level of the lesion
Test reflexes
Deep tendon reflexes
Bulbocavernosus reflex
Cremasteric reflexes
Anal reflexes
Determine the condition of the skin in the perianal area
Excoriation and redness
Note the presence of cognitive impairment or dementia
Any associated conditions (e.g., prostate hypertrophy)
Ascertain the extent of the patient's hand function and ability to perform transfers and activities of daily living (this is especially important in patients who are to perform self-catheterization)

Investigations

Urinalysis and urine culture to rule out infection
Twenty four-hour creatinine clearance
Residual urine volume

Radiologic Studies

X-rays

Plain film of the urinary tract, bladder, and kidneys to determine presence of radiopaque calculi in conjunction with ultrasonography.

Excretory urography or intravenous pyelography can be used for visualization of the collecting system.

Ultrasonography

Urodynamics

Cystometry

Evaluates filling and storage phases of detrusor function by measuring changes in intravesical pressure with increases in bladder volume

Electromyography

Electromyography (EMG) is used to measure electrical potentials generated by depolarization of the detrusor muscle and urethral sphincter.

Cystoscopy

Indicated in patients with recurrent urinary tract infections, bladder stones.

Treatment

A variety of techniques are used to maintain continence and/or empty the bladder.

Goals for Management of the Neurogenic Bladder

Protect the kidneys from hydronephrosis and infection by reducing residual urine and reflux up the ureters

Relieve incontinence

Maintain an acceptable functional capacity so voiding occurs only q4–6h

Valsalva or Credé Maneuver

The Credé maneuver is manual compression of the bladder, used in patients with decreased bladder tone or areflexia and low outlet resistance. Increasing intravesical pressure also may be achieved through the Valsalva maneuver (i.e., abdominal straining).

Initiation of Reflex Bladder Contraction

Pinching or stimulating the lumbar and sacral dermatomal levels is used to provoke reflex bladder contraction. Patients with SCI may use this technique if there is no outlet obstruction or detrusor sphincter dyssynergia.

Timed Voiding

A program of timed voiding is useful in patients with weak sphincters or in patients with hyperreflexic bladders. These patients are put on a schedule of frequent bladder emptying before actual bladder contraction. Timed voiding should be scheduled every 2–4 hours.

Clean Intermittent Catheterization

The practice of clean intermittent catheterization (CIC) is used primarily in patients with neurogenic bladder disease such as in cases of SCI. Prerequisites for use include sufficient outflow resistance to maintain continence between catheterizations, bladder with low pressure, and adequate bladder capacity, ideally more than 300 mL.

Encourage fluid restriction to limit bladder volumes to less than 600 mL.

Limiting Factors

Ataxia

Upper extremity weakness

Excessive adduction spasticity in the legs

Dementia

Complications

Urethral trauma

Predisposition to bacteriuria and/or urinary tract infections.

Indwelling Catheters

Indwelling catheters are either suprapubic or urethral. It is an option for persons who are unable to catheterize themselves.

Surgical Intervention

Surgery on the bladder outlet

Transurethral resection of the bladder neck is indicated in patients with obstruction at the bladder neck when medical therapy has failed to produce satisfactory results.

External sphincterotomy is indicated in patients with suprasacral lesions causing failure to empty when other therapeutic modalities have not been successful. Candidates for this procedure should have adequate detrusor contractions.

Stenting makes use of removable stents inserted into the urethra via cystoscopy. Indications are similar to those for sphincterectomy.

Urethral overdilation is performed only in females and has the same effective as sphincterotomy.

External compressive procedure involves creation of a fascial sling around the bladder neck, using a fascial strip from either the abdominal rectus muscle or tensor fascia lata.
Implantation of an artificial sphincter is used most commonly in children with myelomeningocele who have an incompetent sphincter mechanism.

Surgery on the Bladder

Bladder augmentation is used primarily in patients with refractory hyperreflexic bladder when medical treatment has failed to alleviate symptoms.

Medication (Table 27.2)

Table 27.2 Medications used in bladder dysfunction

Group	Drugs	Uses
Alpha-adrenergic blocking agents	Phenoxybenzamine Prazosin	Useful in reducing bladder outlet resistance Helpful in patients with detrusor sphincter dyssynergia Used to enhance bladder neck resistance in patients with stress incontinence or denervation of the bladder neck Not helpful in areflexic bladders
Anticholinergic agents	Propantheline bromide Oxybutynin Tolterodine tartrate	Urinary incontinence due to uninhibited bladder contractions secondary to suprasacral lesions
Tricyclic antidepressants	Imipramine	Have peripheral alpha-adrenergic and central anticholinergic effects Suppress bladder contractions Enhance bladder neck resistance

Complications

Bladder infections
Pyelonephritis
Kidney stones
If untreated can lead to renal failure

Prognosis

The prognosis for recovery depends on the type, severity, and location of the lesion causing the bladder dysfunction.

BOWEL DYSFUNCTION

Neurogenic bowel dysfunction is common.

Causes of Neurogenic Bowel Dysfunction

Spinal cord injury (SCI)
Amyotrophic lateral sclerosis (ALS)
Spina bifida
Multiple sclerosis (MS)
Diabetes mellitus (DM).

Pathophysiology

Important neural pathways include parasympathetic, sympathetic, and somatic innervation to the colon, rectum, and anus. The vagus nerve innervates the upper segments of the GI tract up to the splenic flexure. The pelvic splanchnic nerves (nervi erigentes) carry parasympathetic fibers from the S2–S4 spinal cord levels to the descending colon and rectum. The hypogastric nerve sends out sympathetic innervation from the L1, L2, and L3 spinal segments to the lower colon, rectum, and sphincters. The pudendal nerve (S2–S4) provides somatic innervation to the external anal sphincter and pelvic floor.

Upper Motor Neurone Lesions

A spinal cord lesion above the conus medullaris
Manifests as underactive propulsive peristalsis, overactive segmental peristalsis, or rectal distention.

Lower Motor Neurone Lesions

A lesion at the level of the conus medullaris, cauda equina, or inferior splanchnic nerve.
Leads to colonic slowing, resulting in constipation, fecal incontinence, and difficulty with emptying.

Clinical Examination

Symptoms of bowel dysfunction may be due to:
Loss of voluntary control over defecation, known as fecal incontinence
Difficulty with evacuation
Associated neurologic bladder symptoms
Associated symptoms of autonomic dysreflexia in patients with spinal cord lesions at T6 and above

Table 27.4 Physical examination

Test	Upper motor neuron	Lower motor neuron
Anal sphincter	Puckered	Flattening or scalloping
Anocutaneous reflex	Present	Absent
Anal tone	Increased	Decreased
Bulbocavernosus reflex	Brisk	Absent

Table 27.4 Differential diagnosis

Neurological causes	Non-neurological causes
Brown-Sequard syndrome Central cord syndrome Multiple sclerosis Myelomeningocele Spinal cord injury Aging	Gastrointestinal neoplasm Gallstone ileus Intestinal adhesions causing obstruction Volvulus Intussusception

Investigations

Endoscopic studies include rectosigmoidoscopy, anoscopy, and colonoscopy to visualize anatomical abnormalities or lesions.

Manometry by kymography and/or catheter use measures pressure and volume changes by intraluminal balloons and catheter, respectively. These tests help determine anorectal pressures and colonic migratory contractions. The saline infusion continence test quantitatively determines continence to liquid after rectal saline infusion.

Electromyography determines the state of innervation of the rectal muscles by their respective motor nerves.

Faecal Impaction

Predisposition Factors for Faecal Impaction

Bedridden patients
Elderly patients, especially those with a history of fecal impaction or constipation.
Patients with abdominal weakness from neuromuscular, neuropathic, spinal cord, or other disorders.
Patients receiving narcotics or other drugs that reduce bowel motility (e.g., anticholinergics)
Dehydrated patients (e.g., those on glycerol, mannitol, or alumina gels)

Management

Adequate hydration
Laxatives
- Natural laxatives (e.g., whole bran, prunes, etc.)
- Dioctyl sulfosuccinate
- Saline laxatives (milk of magnesia (MOM))
- Bisacodyl

Digital removal of the impacted faeces

Fecal Retention and Incontinence

Bowel training
Regular enemas or a suppository
Regular attempts at defecation should be made, using an abdominal corset if necessary.
Maintenance of a soft stool
Adequate hydration
Prunes
Bran cereals
Stool softeners (dioctyl sulfosuccinate)
An abdominal corset may help to increase intra-abdominal pressure in patients with weak abdominal muscles.
Avoid constipating medications, such as narcotics.

Biofeedback

Biofeedback techniques have been used to train the external sphincter and other voluntary muscles to maintain fecal continence.

Surgical Interventions

Sphincteroplasty and gracilis muscle transposition
Continent cecostomy

Section IV

DISORDERS OF PERIPHERAL NERVES

CHAPTER 28

Nerve Injury

An important quality of the peripheral nervous system, as compared to the central nervous system, is its remarkable ability to recover after an injury through remyelination and regeneration of the axon. This chapter will deal in brief with the injuries of the peripheral nerves, approach to these patients and management of these injuries.

CLASSIFICATION AND GRADING OF NERVE INJURY

Seddon (1943), 3 grade (neuropraxia, axonotmesis and neurotmesis) classification scheme was based on clinicopathologic correlation which was further expanded by Sunderland (1951) into 5 degrees of nerve injury. A comparison of two system is shown in Table 28.1.

Table 28.1 Classification of nerve injuries

Seddon	Sunderland
Neuropraxia	Grade I
Axonotmesis	Grade II
	Grade III
	Grade IV
Neurotmesis	Grade V

Neuropraxia

Conduction or complete block of conduction across a segment of a nerve with preservation of axonal continuity

Axonotmesis

Axon continuity is disrupted but with relative sparing of the endoneurium.

Neurotmesis

Endoneurium, perineurium, and epineurium, which make up the entire nerve trunk, are completely divided.

CAUSES OF NERVE INJURY

Blunt forces imparted to nerve remain by far the most common mechanisms of underlying nerve injury. There are several other mechanisms that can lead to nerve injury (Box 28.1)

Box 28.1 Causes of nerve injuries

- Entrapment
- Traction
- Stretch
- Contusion
- Laceration
- Compression and ischemia
- Burns
- Injection and iatrogenic injuries

CLINICAL FEATURES

Pain

Pain is the frequent presenting complaint of patients with peripheral nerve damage.

Sensory Loss

Sensory loss may be complete (anaesthesia) or a decrease (hypoesthesia) in sensation.

Motor Deficit

Patient may have weakness relating to the distribution of nerve (see for details of individual nerve distribution).

Autonomic Dysfunction

Autonomic disturbances may follow the major nerve injuries and it may be a hypofunction of hyperfunction (Box 28.2).

Box 28.2

Autonomic hypofunction
- Loss of sweating
- Cold extremity
- Cyanosis
- Swelling

Autonomic overactivity
- Sympathetically mediated pain syndromes
- Abnormal vasodilatation
- Increased sweating

Physical Examination

General Principles of Peripheral Nerve Exam

- A full exposure of the limb
- Compare one limb to the other
- Systematic and orderly approach, from proximal to distal limb (e.g., parascapular and shoulder girdle muscles before arm and forearm)
- Do examine joint movements (e.g., lateral abduction of the shoulder over the first 30 degrees is produced by supraspinatus, over the next 120 degrees by deltoid and then is completed by medial rotation of the scapula by parascapular muscles such as trapezius and rhomboids).
- Assess and grade individual muscles
- Be aware of and avoid being fooled by trick movements.

Muscle Testing

Compare the involved and the unaffected extremity with respect to motor bulk, tone and then proceed to a thorough evaluation of strength of individual muscle groups.

Sensory Evaluation

Sensory examination includes testing for light touch, pinprick, two-point discrimination, vibration, and proprioception. In complete peripheral nerve injuries all modalities of sensation in the distribution of the nerve will be lost however in incomplete or partial injuries some modalities may be affected more so than others.

Reflexes

Always compare the affected to the unaffected side as myotatic reflexes are extremely sensitive indicators of peripheral nerve pathology.

Autonomic Activity

Inspect the whole limb and digits for autonomic activity and look for:

- Colour
- Temperature
- Sweating behavior (or lack of)
- Atrophic changes in skin organs and nail beds

Tinel Sign

Tinel sign is the finding of shock-like electrical sensations or paresthesias evoked in the nerve distribution by percussing over the injured nerve.

IMAGING STUDIES

Radiography

Many peripheral nerve injuries can be associated with other soft tissue- or bone injuries that can be detected through radiographic findings. For example fracture shaft humerus may be associated with radial nerve injury.

MRI

On conventional MRI signal changes in denervated muscles are seen as early as 4 days after injury and can be better seen with short tau inversion recovery (STIR) sequence. In nerve entrapment and neuropraxic nerve injuries STIR or T2-weighted signals in the innervated muscles remain normal. Also magnetic resonance neurography (MRN) can help visualize both normal and abnormal peripheral nerves in various regions of the body. It is useful in evaluating brachial plexus injuries.

Electrodiagnostic Studies

These objective tests are useful in detecting nerve injury and/or nerve compression and in identifying early stages of recovery.

Electromyography (EMG)

This test is performed at least 4 weeks following nerve injury. EMG testing done prior to that time may yield false-negative findings because it takes 4–6 weeks for muscle fibrillations to become apparent. Evidence of denervation is indicated by the presence of fibrillations in the muscle. Reinnervation is noted by the presence of motor unit potentials.

Nerve Conduction Studies (NCV)

These studies are particularly useful in determining secondary compression sites that may be present. If the nerve is compressed at an entrapment site, such as the carpal tunnel or cubital tunnel, axonal regeneration may be impeded and thus limit reinnervation.

TREATMENT

The goals of treatment in these patients are:

- To return function to the damaged nerve
- To improve the quality of life of patients
- Not only is the nerve treated, but exogenous sources of nerve injury also are treated
- Bone dislocation with neurological deficit requires prompt anatomical reduction to prevent irreversible nerve necrosis
- Analgesics to control pain
- Steroids to decrease endoneurial oedema
- Protection of the joints, including the surrounding ligaments and tendons, from further stress
- Splints, slings, or both

Indications for Surgery

Closed Injuries

If there is no recovery either clinically or with electrodiagnostic studies at 3 months following injury.

Open Injuries (i.e., laceration)

Surgical exploration is recommended as soon as possible.

Crush Injuries

If after three months there is no evidence of reinnervation then surgical reconstruction is recommended.

Grade V

Early surgical repair is recommended.

CHAPTER 29

Peripheral Neuropathies

Peripheral neuropathies can be categorized on the basis of the structure primarily affected. The predominant pathologic feature may be axonal degeneration (axonal or neuronal neuropathies) or paranodal or segmental demyelination. The distinction may be possible on the basis of neurophysiologic findings.

Mononeuropathies

An individual nerve may be injured along its course or may be compressed, angulated, or stretched by neighbouring anatomic structures, especially at a point where it passes through a narrow space (entrapment neuropathy). Mononeuropathies lead to a sensory, motor, or mixed deficit that is restricted to the territory of the affected nerve. A similar clinical disturbance is produced by peripheral nerve tumours, but these are rare except in patients with Recklinghausen's disease.

- Carpal tunnel syndrome
- Pronator teres or anterior interosseous syndrome
- Ulnar nerve lesions
- Radial nerve lesions
- Femoral neuropathy
- Meralgia paresthetica
- Sciatic and common peroneal nerve palsies
- Tarsal tunnel syndrome
- Facial neuropathy
- Bell's palsy

Polyneuropathies

Symmetric loss of sensory function, motor function, or both, usually begin in the feet in polyneuropathy. Deep tendon reflexes may be diminished

early, especially in the ankles; later, reflexes may be absent. Some neuropathies affect the proximal nerves early (diabetic amyotrophy), and these may be confused with myopathy.

Multiple Mononeuropathies (Mononeuritis multiplex)

Multiple mononeuropathies suggest a patchy multifocal disease process.

- Vasculopathy (e.g., diabetes, arteritis)
- Infiltrative process (e.g., leprosy, sarcoidosis)
- Radiation damage
- Immunologic disorder (e.g., brachial plexopathy)

Diffuse Polyneuropathies

Diffuse polyneuropathies lead to a symmetric sensory, motor, or mixed deficit, often most marked distally.

Hereditary

- Charcot-Marie-Tooth disease
- Dejerine-Sottas disease (HMSN Type III)
- Friedreich's ataxia
- Refsum's disease (HMSN Type IV)
- Porphyria

Systemic and Metabolic

- Diabetes mellitus
- Amyloidosis
- Uraemia
- Myxoedema
- Acromegaly
- Alcoholism
- Nutritional deficiency (beri beri, pyridoxine deficiency, vitamin B_{12} deficiency)
- Paraproteinemias

Toxins

- Acrylamide
- Arsenic
- Lead
- Mercury
- Thallium
- Carbon disulphide

Iatrogenic

Cisplatin

- Isoniazid
- Nitrofurantoin
- Vincristine
- Dapsone
- Disulfiram

Infectious & Inflammatory Diseases

- Leprosy
- AIDS
- Lyme borreliosis
- Sarcoidosis
- Polyarteritis
- Rheumatoid arthritis
- Idiopathic inflammatory polyneuropathy (Guillain-Barre syndrome)
- Chronic inflammatory polyneuropathy

Neuropathies Associated with Malignant Diseases

Neuropathy Associated with Critical Illness

Patients in intensive care units with sepsis and multiorgan failure sometimes develop polyneuropathies.

INVESTIGATIONS

The first step in management of a neuropathy is to diagnose the underlying disease or cause. Investigations in peripheral neuropathy should be ordered selectively, as guided by symptoms and signs (Box).

Complete blood count
Erythrocyte sedimentation rate
Serum protein electrophoresis
Blood urea and serum electrolytes
Liver and thyroid function tests
Rheumatoid factor and antinuclear antibody
HBsAg determination
Serologic tests for syphilis
Fasting blood glucose level
Urinary heavy metal levels
Cerebrospinal fluid examination
Chest radiography

Nerve conduction velocity is important in confirming the peripheral nerve origin of symptoms and providing a means of following clinical changes, as well as indicating the likely disease process (i.e., axonal or demyelinating neuropathy).

Cutaneous nerve biopsy may help establish a precise diagnosis (e.g., polyarteritis, amyloidosis).

PRINCIPLES OF TREATMENT

Treatment is of the underlying cause, when feasible.
Physical therapy helps prevent contractures
Splints can maintain a weak extremity in a position of useful function
Anaesthetic extremities must be protected against burns, injury
Shoes should be examined frequently during the day for grit or foreign objects in order to prevent pressure lesions.

Neuropathic Pain (management is discussed in detail in Chapter 43)

Analgesics (aspirin)
Narcotics or narcotic substitutes for severe hyperpathia or pain
Phenytoin, carbamazepine, gabapentin, or tricyclic antidepressants may be needed.

Postural Hypotension

Wearing waist-high elastic stockings and sleeping in a semierect position at night.
Fludrocortisone
Midodrine

ACUTE IDIOPATHIC POLYNEUROPATHY (GUILLAIN-BARRE SYNDROME)

This acute or subacute polyradiculoneuropathy sometimes follows infective illness, inoculations, or surgical procedures. There is an association with preceding Campylobacter jejuni enteritis.

Clinical Features

The earliest signs are motor weakness, paresthesias, and pain, progressing from lower extremities to upper extremities.

The cranial nerves are affected in 75% of the cases. The facial nerve is involved in half of those with cranial neuropathy, and of those with facial palsies. It is bilateral in about 80% cases.

Autonomic disorders (cardiac arrhythmia, hypotension, hypertension, hyperpyrexia, and tachycardia) occur frequently and account for almost half of fatalities.

Investigations

The cerebrospinal fluid characteristically contains a high protein concentration with normal cell content, but these changes may take 2 or 3 weeks to develop.

Nerve conduction velocities are reduced in 90% of patients.

Differential Diagnosis

Porphyric
Diphtheritic
Toxic (heavy metal, hexacarbon, organophosphate) neuropathies
Poliomyelitis
Botulism
Tick paralysis

Treatment

Prednisone is ineffective and may prolong recovery time.
Plasmapheresis is best performed within the first few days of illness and is best reserved for clinically severe or rapidly progressive cases or those with ventilatory impairment. Intravenous immunoglobulin.

Respiratory Care

The most important consideration is maintaining adequate respiration. Patients should be followed closely, with measurement of vital capacity, and endotracheal intubation is performed when the vital capacity drops to 25% to 30% of normal.
When bulbar muscles are involved, feeding is done by intravenous administration, nasogastric tube, or gastrostomy.
Prophylactic anticoagulation is indicated in paralyzed patients.
Fecal impaction is a painful problem that occurs in bedridden patients. Thus, suppositories and enemas should be used from the outset, before problems arise.

Physical Therapy

Bed rest is indicated in the acute phase of the illness until improvement begins.
Prevention of contractures.
Careful positioning of the patient in bed prevents bed sores and compression of nerves.

Pain

Mild analgesics (aspirin, 600 mg q3–4h), but occasionally codeine (30 to 60 mg q4h) or morphine (5 to 10 mg IM) is needed. Narcotics cause respiratory depression and constipation.

Autonomic Dysfunctions

Occasionally, paroxysmal hypertension, headache, sweating, anxiety, and fever occur in patients with acute polyradiculoneuritis and these have to be managed accordingly.

Prognosis

Most patients eventually make a good recovery, but this may take many months, and 10–20% patients are left with persisting disability. Approximately 3% of the patients with acute idiopathic polyneuropathy have one or more clinically similar relapses, sometimes several years after the initial illness. Plasma exchange therapy may produce improvement in chronic and relapsing inflammatory polyneuropathy.

DIABETIC NEUROPATHY

A significant degree of peripheral neuropathy develops in about 15% of the patients with diabetes.

Pathology

The neuropathies that appear can be divided into symmetrical, sensory, polyneuropathies, and autonomic neuropathies, and isolated, peripheral, nerve lesions, or multifocal neuropathies. Isolated nerve lesions occur more commonly in elderly diabetic subjects.

Clinical Features

The commonest form is a mild symmetrical, sensory, polyneuropathy, giving rise to numbness and tingling paraesthesiae in the toes and feet and less often in the fingers.
Examination reveals loss of vibration sense in the feet, depression of the ankle jerks, and mild distal cutaneous sensory impairment.
Autonomic neuropathy frequently accompanies the sensory neuropathy and may be the salient manifestation. Symptoms of autonomic neuropathy include dysphagia from oesophageal involvement, episodes of vomiting related to gastric atony, and episodic nocturnal diarrhoea, often alternating with periods of constipation. Those related to the genitourinary system include impotence, retrograde ejaculation, and bladder atony with difficulty in voiding and urinary retention with overflow. Vascular denervation results in postural hypotension, and cardiac denervation may be demonstrable by an elevated resting heart rate and the absence of beat-to-beat variation with respiration.

Treatment

Control of Diabetes

Care of the feet in diabetic sensory neuropathy, to prevent the development of chronic ulceration.

Control of Pain

Carbamazepine, tricyclic antidepressants

Postural Hypotension

Postural hypotension can be improved by raising the head of the bed at night or by support bandages to the legs; more severe cases may require treatment with fludrocortisone.

Gastroparesis

Gastroparesis may respond to metoclopramide, domperidone, or erythromycin.

Diarrhoea

Diabetic diarrhoea can be helped by low-dosage tetracycline or diphenoxylate, loperamide, or codeine phosphate.

CARPAL TUNNEL SYNDROME

Carpal tunnel syndrome is the most common entrapment neuropathy. It is the result of compression of the median nerve by the volar ligament.

Clinical Features

Pain and tingling in the hand are the usual early signs, and retrograde distribution of the pain may cause arm and shoulder pain in some patients. There may be no objective neurologic signs, and nerve conduction studies may be normal, but usually median nerve conduction is slowed across the wrist. Often, the earliest objective sign is failure to appreciate textures. Later, clear deficits of sensation with muscle wasting occur in the distribution of the median nerve.

Treatment

The treatment is usually surgical release at the site of entrapment.
When the symptoms are mild and there are no objective signs of nerve damage, conservative measures may be enough. For example, splinting the wrist at night may relieve carpal tunnel symptoms, especially in the transient syndrome seen in pregnancy.

Injection of corticosteroid into the volar ligament may relieve the symptoms temporarily. In some cases, remission may last for several years. If nerve conduction is slow, however, surgery is usually required.

CHAPTER 30

Peripheral Nerve Tumours

Peripheral nerve tumours can be localized (neurilemmoma) or diffusely invasive (plexiform neurofibroma). These tumours can present in many locations and the clinical presentation will vary accordingly.

PATHOLOGY (BOX 30.1)

The most common of the type of nerve sheath tumours is neurilemmoma, or schwannoma. Neurilemmomas are benign encapsulated tumours of the nerve sheath and their cell of origin is thought to be Schwann cells derived from the neural crest. These masses usually arise from the side of a nerve and are well encapsulated. Neurofibromas are most often found in patients with neurofibromatosis. Rarely, these tumours may become malignant, metastasizing to other portions of the body and invading surrounding tissues.

Box 30.1 Histological types

- Neurilemmoma or schwannoma
- Fibroma
- Neurofibroma
- Neurosarcoma
- Ganglion cyst
- Giant cell tumour of tendon sheath
- Lipoma

CLINICAL FINDINGS

The symptoms and signs are those of peripheral nerve dysfunction and related to a specific nerve rather than tracts in the spinal cord or brain. Clinical symptoms can be irritative (pain) or paralytic (weakness of muscles).

Typically these tumours are slow growing and can be present for months to years without symptoms.
The head and flexor surface of the upper and lower extremities and the trunk are common locations in decreasing order.
Presentation can be either an asymptomatic mass or mild localized pain or paresthesia due to pressure on the nerve of origin.
There may be local tenderness on palpation.
The mass is usually mobile in the transverse plane and tethered along the axis of the nerve from which it arises.
Tumour occurring in well-defined compartments (e.g., wrist, ankle), they can present as carpal tunnel syndrome or tarsal tunnel syndrome.
Lesions in the sciatic nerve can mimic discogenic low back pain.

INVESTIGATIONS

Nerve conduction tests and electromyography – to localize the nerve dysfunction

MRI

MRI is particularly useful and shows a usually round or oval mass with a moderately bright signal on T1-weighted images and a bright heterogeneous signal on T2-weighted images. The lesion enhances uniformly with gadolinium contrast.

DIFFERENTIAL DIAGNOSIS

Peripheral neuropathies

TREATMENT

Surgery

The operative approach to peripheral nerve tumours depends on the type of lesion.
As with most benign tumours, neurilemmomas respond well to local resection and complete resection is curative in these benign lesions.

Complications

The most common complication is initial neuropraxia; however, this neurologic deficit can be permanent depending on the resection of neural tissue.

PROGNOSIS

Recurrence is unlikely with complete resection. Malignant change is

Section V

DISORDERS OF MUSCLES AND NEUROMUSCULAR JUNCTION

CHAPTER 31

Myopathic Disorders

MYOPATHIC DISORDERS

- Muscular dystrophies
- Metabolic and endocrine myopathy
- Inflammatory myopathy or polymyositis
- Congenital myopathy
- Toxic myopathy

MUSCULAR DYSTROPHY

Muscular dystrophy is a group of congenital disorders characterized by progressive symmetric wasting of skeletal muscles without neural or sensory defects. Paradoxically, some wasted muscles tend to enlarge (pseudohypertrophy) because connective tissue and fat replace muscle tissue, giving a false impression of increased muscle strength.

Main types of muscular dystrophy are:
- Duchenne, or pseudohypertrophic; 50% of all cases
- Becker, or benign pseudohypertrophic
- Facioscapulohumeral, or Landouzy-Dejerine dystrophy
- Limb-girdle dystrophy

Clinical Features (Table 31.1)

Duchenne Muscular Dystrophy

Insidious onset between the ages of 3 and 5 years
Initial effect on legs, pelvis, and shoulders
Waddling gait, toe-walking, and lumbar lordosis due to muscle weakness
Difficulty in climbing stairs, frequent falls
Enlarged, firm calf muscles

[illegible] to wheelchair (usually by 9 to 12 years of age)

[illegible]ker (benign pseudohypertrophic) Muscular Dystrophy

[illegible]ilar to those of Duchenne muscular dystrophy but with slower progression

[illegible]cioscapulohumeral (Landouzy-Dejerine) Dystrophy

[illegible]kness of face, shoulder, and upper arm muscles (initial sign)
[illegible]dulous lip and absent nasolabial fold
[illegible]bility to pucker mouth or whistle
[illegible]normal facial movements and absence of facial movements when laughing [illegible]ying
[illegible]lose facial flattening leading to a mask-like expression
[illegible]bility to raise arms above the head

[illegible]mb-girdle Dystrophy

[illegible]kness in upper arms and pelvis first
[illegible]mbar lordosis with abdominal protrusion
[illegible]nging of the scapulae
[illegible]ddling gait
[illegible] balance
[illegible]bility to raise the arms

[illegible]ble 31.1 Salient features of muscular dystrophies

Type of dystrophy	Chromosome link	Inheritance	Age at onset
[illegible]uchenne, or [illegible]eudohypertrophic	X	X-linked recessive	3–5
[illegible]ecker, or benign [illegible]seudohypertrophic	Multiple	X-linked recessive	3–5
[illegible]cioscapulohumeral, [illegible] Landouzy-Dejerine, [illegible]ystrophy	4	Autosomal dominant disorder	10–40
[illegible]mb-girdle dystrophy	19	Autosomal recessive disorder	10–30

[illegible]mplications

[illegible]diac and respiratory muscle weakness leading to tachycardia, [illegible]trocardiographic abnormalities, and pulmonary complications. Death [illegible]monly is due to sudden heart failure, respiratory failure, or infection.

Diagnosis

Diagnosis depends on typical clinical findings, family history, and diagnostic test findings. If another family member has muscular dystrophy, its clinical characteristics can suggest the type of dystrophy the patient has and how he may be affected.

Investigations

Electromyography

Electromyography showing short, weak bursts of electrical activity in affected muscles

Muscle Biopsy

Muscle biopsy showing a combination of muscle cell degeneration and regeneration (in later stages, showing fat and connective tissue deposits)

Creatine Kinase

Serum creatine kinase is markedly elevated in Duchenne and Becker dystrophies.

Immunologic and Molecular Biological Techniques

Immunologic and molecular biological techniques facilitate accurate prenatal and postnatal diagnosis of Duchenne and Becker dystrophies.

Treatment

No specific treatment is available to stop the progression of disease and muscle impairment.

Supportive Treatments

Coughing and deep-breathing exercises and diaphragmatic breathing
Orthopaedic appliances, exercise, physical therapy, and surgery to correct contractures (to help preserve mobility and independence)
Genetic counseling regarding risk of transmitting disease for family members who are carriers
Adequate fluid intake, increased dietary bulk, and stool softener for constipation due to inactivity
Low-calorie, high-protein, high-fiber diet (physical inactivity predisposes to obesity).

CHAPTER 32

Myasthenia Gravis

Myasthenia gravis causes progressive weakness and abnormal fatigability of striated (skeletal) muscles; symptoms are exacerbated by exercise and repeated movement and relieved by anticholinesterase drugs. Commonly it affects muscles innervated by the cranial nerves (face, lips, tongue, neck, and throat), but it can affect any muscle group.

CAUSES

The exact cause of myasthenia gravis is unknown. However, it is believed to be the result of:
Autoimmune response
Ineffective acetylcholine release
Inadequate muscle fiber response to acetylcholine

PATHOLOGY

Myasthenia gravis causes a failure in transmission of nerve impulses at the neuromuscular junction. The site of action is the postsynaptic membrane. Theoretically, antireceptor antibodies block, weaken or reduce the number of acetylcholine receptors available at each neuromuscular junction and thereby impair muscle depolarization necessary for movement.

CLINICAL FEATURES

Myasthenia gravis may occur gradually or suddenly. Progressive muscle weakness and accompanying loss of function depending on muscle group affected; becoming more intense during menses and after emotional stress, prolonged exposure to sunlight or cold, or infections
Its signs and symptoms include the following:
Difficulty chewing and swallowing

Facial and Extraocular Muscles

Weak eye closure
Ptosis
Diplopia
Blank and expressionless facial appearance

Lower Cranial Nerve Weakness

Nasal vocal tones
Frequent nasal regurgitation of fluids

Skeletal Muscles

Skeletal muscle weakness and fatigue, increasing through the day but decreasing with rest, in late stages it may be severe enough to cause paralysis.
Neck muscles weakness with head tilting back.
Respiratory muscles weakness, decreased tidal volume and vital capacity from impaired transmission to the diaphragm (predisposing to pneumonia and other respiratory tract infections).
Clinical examination will confirm the weakness and fatigability of affected muscles.

COMPLICATIONS

Respiratory distress
Pneumonia
Aspiration
Myasthenic crisis

DIAGNOSIS

Tensilon Test

Tensilon test confirms diagnosis of myasthenia gravis, revealing temporarily improved muscle function within 30 to 60 seconds after IV injection of edrophonium or neostigmine and lasting up to 30 minutes.

Electromyography

Electromyography with repeated neural stimulation shows progressive decrease in muscle fiber contraction.

Serum Antiacetylcholine Antibody

Serum antiacetylcholine antibody titer may be elevated.

Imaging

- Lateral and anteroposterior X-rays of the chest and CT scans to demonstrate pre-existing thymoma.

TREATMENT

Anticholinesterase Drugs

- Anticholinesterase drugs (neostigmine, pyridostigmine) to counteract fatigue and muscle weakness, and allow about 80% of normal muscle function.

Immunosuppressant Therapy

- Corticosteroids
- Azathioprine
- Cyclosporine
- Cyclophosphamide
- IVIG during acute relapses
- Plasmapheresis in severe exacerbations to suppress the immune system
- Thymectomy to remove thymomas and possibly induce remission in some cases of adult-onset myasthenia

Supportive Measures

- Tracheostomy, positive-pressure ventilation, and vigorous suctioning to remove secretions for treatment of acute exacerbations that causes severe respiratory distress.
- Continuation of anticholinesterase drugs in myasthenic crisis, until respiratory function improves (Myasthenic crisis requires immediate hospitalization and vigorous respiratory support).

PROGNOSIS

Myasthenia gravis follows an unpredictable course of periodic exacerbations and remissions. There is no known cure. Drug treatment has improved the prognosis and allows patients to lead relatively normal lives, except during exacerbations. Though the disorder follows a slowly progressive course, it rarely have a fatal outcome owing to respiratory complications such as aspiration pneumonia.

CHAPTER 33

Polymyositis

Polymyositis is an inflammatory autoimmune muscle disease that causes muscular weakness and wasting and may be associated with muscle pain and tenderness or with evidence of some form of connective tissue or collagen disease.

CLINICAL FEATURES

Clinical features are variable but proximal muscle weakness is the rule. Usually disease is subacute (course that evolves over several months) on onset but in some cases it may be acute and may progress in weeks to complete disability.

Face is often spared, and muscle atrophy is a late sign

Pharyngeal muscle involvement may produce dysphagia

Speech usually remains normal

Posterior neck muscles are often weak.

Some patients develop an erythematous skin eruption on the dorsum of the hands, proximal digits, knees, or elbows, or a purple discolouration and oedema of the eyelids (dermatomyositis).

Cardiac involvement occurs in up to 30% of patients.

Muscle pain and tenderness

Joint pain and stiffness

DIAGNOSIS

Muscle Enzymes

Muscle enzymes (CK, transaminase, and aldolase) are usually elevated, but they may be normal in some cases.

ESR

The erythrocyte sedimentation rate does not correlate reliably with the activity of the disease.

EMG

The EMG is nonspecific, but it usually suggests a primary myopathic process with brief, small-amplitude polyphasic motor units.

Muscle Biopsy

Muscle biopsy shows an inflammatory infiltrate and necrosis of muscle fibers in about 85% to 90% of patients.

DIFFERENTIAL DIAGNOSIS

Paroxysmal myoglobinuria

Parasitic myositis

Polymyalgia rheumatica

Muscular dystrophy

Thyrotoxic and other endocrine myopathies

Diabetic amyotrophy.

TREATMENT

Corticosteroids

Corticosteroids have been found to benefit patients with polymyositis. Corticosteroids are usually used in combination with either methotrexate or azathioprine.

Immunotherapy

Immunosuppressive drugs (Methotrexate, Azathioprine) are indicated in patients with chronic progressive polymyositis, not improved by corticosteroid therapy, and in those who have intolerable complications of steroid therapy.

Intravenous Immunoglobulin G (IVIG)

Intravenous immunoglobulin G (IVIG) has been used in some cases of polymyositis but there is not yet convincing evidence that it is better than corticosteroids.

Supportive Measures

Bed rest in acute cases.

Physical therapy is indicated for range-of-motion exercises.

Patients with chronic weakness may benefit from braces and other physical measures.

PROGNOSIS

The natural history of the disease varies, relapsing and remitting when treated and occasionally improving spontaneously. Cancer, cardiac involvement, older age of onset, and delay of treatment are each associated with a poor prognosis. Other organ systems are also involved in polymyositis. Interstitial lung disease is one of the most common complications.

Section VI

OTHER COMMON CONDITIONS

CHAPTER 34

Neurodevelopmental Disorders

Human brain displays plasticity, meaning that with specific stimulation, function, structure and even chemistry of the brain and central nervous system changes when impacted specifically by stimulation. This means that human function, which is controlled by the central nervous system and more specifically the brain, is changeable. If one can evaluate what is causing problems in development and can find the specific stimulation that can impact that development, one can accelerate the development and help improve function.

DEVELOPMENTAL DOMAINS

It's vital to look at any dissociation between the domains of development. The most common developmental domains are as follows:

- Global
- Speech and language
- Motor
- Fine motor
- Personal and social

DEVELOPMENTAL DELAY

To be developmentally delayed means that in some way, a child is functioning at least one to two years behind in areas of cognition, speech and language, gross and/or fine motor areas. When evaluating delayed development, it's important to consider three processes: delay, dissociation and deviancy.

Delay

Delay refers to a significant lag in one or more areas of development.

Dissociation

Dissociation is the difference between the developmental rates of two domains—one being more delayed.

Deviancy

Deviancy refers to nonsequential unevenness in achieving milestones in one or more domains of development.

GLOBAL DEVELOPMENTAL DELAY

Children with a diagnosis of mental retardation often present with mixed or global developmental delays. Common reasons for global developmental delay include chromosomal anomalies (e.g., Down syndrome), fetal alcohol and fragile X syndrome.

SPEECH AND LANGUAGE DELAY

Disorders of speech and language development are most prevalent in children with developmental disabilities.

EXPRESSIVE LANGUAGE DELAY

Expressive language delay is the most common developmental presentation in a primary-care setting. The social and educational development of children with delayed speech and language may be significant. Clinical diagnoses to consider when a child presents with delayed speech and language include hearing loss, mental retardation, autism, dysarthria, a specific learning disability, and developmental language disorders.

HEARING LOSS

All children with delayed speech and language should have an audiometric assessment. Congenital sensorineural hearing loss may cause delayed speech and language. Language skills are more affected by bilateral sensorineural hearing loss and the greater the hearing loss, the greater the language deficits.

MENTAL RETARDATION

Children with delayed speech and language should be evaluated for cognitive abilities. There's a close association among social and affective abilities, and cognitive, sensory, and language development.

AUTISM

Autism is one of the most complex neurodevelopmental disorders. Children with autism have significant communication impairment.

DYSARTHRIA

Oral motor dysfunction of the speech-producing musculature (in which children have dysarthria or mechanical difficulties in speaking) is seen in children with cerebral palsy and other conditions. It leads to uncoordinated oral musculature. It is important to rule out a cleft lip and palate or malformation of the velopharyngeal part in these children.

VERBAL LEARNING DISABILITY

A verbal learning disability is often associated with speech and language. Children with a specific learning disability—similar to children with severe mental retardation or autism—may present with dissociation in developmental skills. Their language may be more delayed than their motor skills.

DEVELOPMENTAL LANGUAGE DISORDERS

In a developmental language disorder, impaired language can't be attributed to a neurological or general medical condition. It's characterized by a slow rate of language development, in which speech begins late and advances slowly.

MOTOR DELAY

Motor delay is most common during the first six to 18 months of a child's life. Early motor delays are often a sign of neurological dysfunction (Box 34.1). When a motor delay is combined with delays in other developmental domains, examining the child for visual impairment or a mental handicap should be considered. Older children with poor motor skills may have a development coordination disorder where their motor skills are substantially below their cognitive abilities. Their clumsiness may be associated with a learning disability or attention-deficit hyperactivity disorder.

Box 34.1

Motor delay
Cerebral palsy
Ataxia
Spina bifida
Spinal muscular atrophy
Myopathy
Mental retardation
Visually impairement

FINE-MOTOR ADAPTIVE DELAY

Hemiplegia
Erb's palsy
Klumpke's paralysis
Visual impairment
Mental retardation
Developmental coordination disorder

CHAPTER 35

Treatment Approaches

Theoretical approaches that serve as the basis of treatment for patients with lesions affecting the nervous system have been based on continually evolving understanding of neurophysiology. These traditional theories incorporate abnormal movement patterns, manual techniques, or both to inhibit (to hold back or keep from some action) or facilitate (to make easy or easier) movement. For the treatment purposes patients can be divided into two groups, low-functioning and high functioning. Low-functioning patients require manual assistance to perform functional movement. They may benefit from the application of selected facilitation techniques to promote motor activity in a posture or during a functional movement. High-functioning patients are capable of performing the tasks, but their movement strategies may be inefficient, unsafe, or both.

THE TRADITIONAL THEORETICAL APPROACHES

The traditional theoretical approaches described are based on the work of Margaret Rood; Berta and Karl Bobath (NDT); Signe Brunnstrom; and Herman Kabat, Margaret Knott, and Dorothy Voss (PNF).

ROOD' TECHNIQUE

This treatment approach to the neurologically impaired patients covered two major areas:
1) Motor development, or designs of movement
2) Sensory stimulation techniques

According to Rood, the sequence of motor development encompasses these four concepts: mobility, stability, controlled mobility, and skill.

Rood's Four Stages of Motor Development

Mobility

Free, flexible movement that encompasses qualities of range and speed

Stability

Motor function that fixes the body to enable weight-bearing and later in the motor progression enables dynamic holding during movement

Controlled Mobility

Creation of the distal segment of an extremity with movement of the proximal segment. An example of this is rocking forward and backward in the quadruped position. The hands and knees are fixed in weight-bearing positions while the shoulder, pelvis, and trunk move.

Skill

Coordinated movement that enables the distal segment of an extremity to manipulate an object while in a stabilized posture. In the quadruped position, skill is demonstrated by reaching up with one upper extremity to explore the environment as the opposite upper extremity provides stability. In Rood's technique sensory stimuli are used to facilitate or inhibit responses. These sensory stimuli can be categorized either facilitatory or inhibitory.

Facilitation

- Approximation: Drawing the bones of a joint together (the application of pressure to a joint)
- Ice applications (3–5 seconds): Quick stroking of an ice cube on the skin.
- Resistance: A strengthening exercise done by applying an outside force to a muscle to force it to develop greater tension.
- Tapping: The tapping of the muscle belly of weak muscles to produce volitional contraction.
- Traction: Drawing or pulling of the joints of spine or extremities.
- Joint compression: The application of pressure to a joint.
- Light touch: The application of a gentle touch, such as with a cotton swab.
- Quick stretch: A quick elongation of a muscle at its lengthened state or during a contraction.

Inhibition

- Prolonged stretch or deep pressure
- Warm or neutral temperature
- Carotid reflex

- Prolonged cold (15–20 minutes)
- Slow stroking down posterior rami

Once the desired response is obtained (i.e., muscle activation or inhibition), the sensory stimulus should be withdrawn as repeated application of the stimulus may cause conditioned response to the stimulus. Facilitation techniques may be beneficial in assisting muscle activation during a functional task.

KARL AND BERTA BOBATH: NEURODEVELOPMENTAL TREATMENT (NDT)

Karl and Berta Bobath observed that abnormal tone and coordination problems (or muscular imbalances) were due to the release of abnormal postural reflexes (primitive reflexes) seen at brain stem or spinal cord levels that inhibited righting reactions, equilibrium reactions, and automatic movements. Abnormal patterns of posture and movement resulted from the loss of CNS control as a consequence of disease or injury.

The goal of NDT is to "achieve a balance between muscle groups and to decrease the effects of abnormal tonal influence on automatic responses and movement patterns". NDT treatment emphasized normalizing muscle tone, inhibiting primitive reflexes, and facilitating normal postural reactions through the developmental sequence.

Treatment Principles

- To change abnormal patterns of movement with dynamic reflex inhibiting patterns (RIPs). The RIPs are used to move the extremities out of the abnormal positions that developed from abnormal tone and into antagonistic patterns. For example, an arm that is flexed and internally rotated is extended and externally rotated. The RIP decreases abnormal tone in flexor components, while dynamically positioning the extremity for the activation of extensor muscles.
- To use key points of control (neck and spine, shoulders, pelvis, toes and ankles, and fingers and wrists) as manual contacts.
- To replace abnormal tone or patterns of movement immediately with normal movement patterns.
- Varying the activity level according to the level of difficulty the patient can handle.
- Varying the context in which the activity occurs.

Neurodevelopmental treatment has evolved over the past several years to incorporate activities that encourage functional carryover. Most activities used in NDT are functional activities that patients performed before onset of their disability.

BRUNNSTROM

Brunnstrom's theory was based on the hierarchical model developed by Hughlings Jackson. Signe Brunnstrom identified seven stages of recovery, based on observing her patients after a stroke (Box). The seven recovery stages described by Brunnstrom are used extensively to document motor recovery seen in patients with stroke.

Box 35.1 Brunnstrom's motor stages

Stage I	Flaccid stage: No muscle tone can be sensed
Stage II	Spastic stage: Presence of muscle spasticity or associated movement
Stage III	Synergy stage: Presence of stereotype of motor synergy
Stage IV	Movement deviating from the basic synergies: Begin to break stereotyped of motor synergy
Stage V	Relative independence of the basic synergies Selective movement of different joints is adequate
Stage VI	Near normal stages
Stage VII	Recovery stage, normal motor function is restored

Brunnstrom advocated the use of cutaneous and muscle sensations and maximally resisting voluntary movement of normally innervated muscles to create overflow to recruit the involved musculature. For example, tapping on the biceps brachii muscle belly can be used to activate cutaneous receptors and muscle spindles to increase the contractile response from the muscle.

KABAT, KNOTT, AND VOSS: PROPRIOCEPTIVE NEUROMUSCULAR FACILITATION (PNF)

The goal of proprioceptive neuromuscular facilitation technique is to strengthen muscles in the movement patterns in which they are designed to function. The patterns of motion used in PNF are mass movement patterns, which are characteristic of normal motor activity. These patterns are spiral and diagonal, in keeping with the spiral and rotary characteristics of skeletal muscle, and closely resemble the movements used in sports and work activities. The diagonal patterns are designed to address specific problems, such as weakness through partial ranges, lack of stability, and weakness in eccentric contractions. Manual contacts used in PNF facilitate underlying muscles and are used to apply resistance in the movement pattern to activate muscle spindles or Golgi tendon organs.

Treatment Techniques

- Slow reversal and slow reversal hold
- Repeated contractions with quick stretch
- Agonistic reversal
- Rhythmic initiation
- Hold relax active movement
- Hold relax or contract relax
- Rhythmic stabilization

TASK ORIENTED MODEL

The systems approach to treatment is geared primarily to task-oriented activities inclusive of the interaction of multiple physiologic systems (i.e., muscles, joints, CNS), the environment, and the individual's motivation to complete the task. In the systems model, no one system dominates another (as is characteristic of the hierarchical model).

CHAPTER 36

Motor Control and Learning

THE PRIMARY MOTOR CORTEX (BRODMANN'S AREA 4)

The primary motor cortex lies along the precentral gyrus, and generates the signals that control the execution of movement. The primary motor cortex is located in the frontal lobe of the brain, along the precentral gyrus. The role of the primary motor cortex is to generate neural impulses that control the execution of movement. Signals from primary motor cortex cross the midline to activate skeletal muscles on the opposite side of the body, meaning that the left hemisphere of the brain controls the right side of the body, and the right hemisphere controls the left side of the body.

Motor Homunculus

Every part of the body is represented in the primary motor cortex, and these representations are arranged somatotopically. The cortical representation of each body part is proportionate in size to the skill with which the part is used in fine, voluntary movement. The areas involved in speech and hand movements are larger than other areas.

SECONDARY MOTOR AREAS

Secondary motor areas are involved in motor planning and include posterior parietal cortex, the premotor cortex, and the supplementary motor area.

The posterior Parietal Cortex

The posterior parietal cortex is involved in transforming visual information into motor commands. Lesions of the somatic sensory area cause deficits in motor performance that are characterized by inability to execute learned sequences of movements such as eating with a knife and fork.

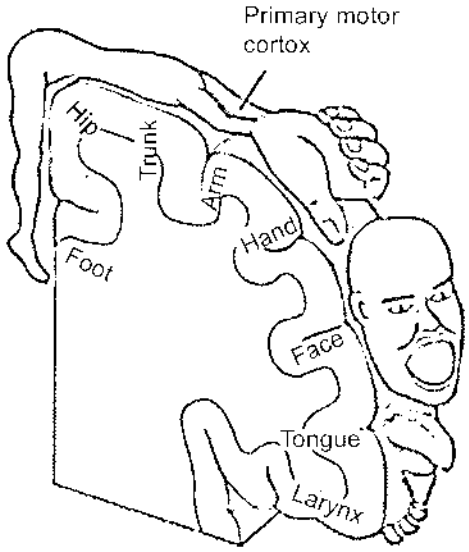


Fig. 36.1 The motor homunculus in primary motor cortex

The Premotor Cortex

The premotor cortex lies just in front of (anterior to) the primary motor cortex. It is involved in the sensory guidance of movement, and controls the more proximal muscles and trunk muscles of the body.

The Supplementary Motor Area

The supplementary motor area lies above, or medial to the premotor area, also in front of the primary motor cortex. It is involved in the planning of complex movements and in coordinating two-handed movements. The supplementary motor area and the premotor regions, both send information to the primary motor cortex as well as to brainstem motor regions.

CORTICOSPINAL TRACT

Neurons in primary motor areas, sensory motor areas and premotor cortex give rise to the fibers of the corticospinal tract. The corticospinal tract is the only direct pathway from the cortex to the spine and is composed of over a million fibers. These fibers descend through the brainstem where the majority of them cross over to the opposite side of the body. After crossing, the fibers continue to descend through the spine, terminating at the appropriate spinal levels. The corticospinal tract is the main pathway for control of voluntary movement in humans.

EXTRAPYRAMIDAL SYSTEM

There are other motor pathways which originate from subcortical groups of motor neurons (nuclei). These pathways control posture and balance, coarse movements of the proximal muscles, and coordinate head, neck and eye movements in response to visual targets. Subcortical pathways can modify voluntary movement through interneuronal circuits in the spine and through projections to cortical motor regions.

MOTOR LEARNING

The ability to learn defines the requirement to be able to acquire and stabilize new motor programmes. Primary principles that govern human learning include: clear goals, focused practice, corrective feedback and motivation.

THE LEARNING STAGES

Learners appear to go through three phases of practice. The first stage is the cognitive stage where the primary task is to understand what is to be done. Motor skills involve three distinct phases of learning:

- Cognitive
- Associative
- Autonomous

MOTOR SKILLS DISORDER

Motor coordination is the product of a complex set of cognitive and physical processes, often taken for granted in children who are developing normally. Smooth, targeted, and accurate movements, gross and fine, necessitate the harmonious functioning of sensory input, central processing of this information in the brain and coordination with the higher executive cerebral functions (e.g., volition, motivation, motor planning of an activity), and finally, carrying out of a certain motor pattern. These elements must work in a coordinated and rapid way to enable the execution of complex movements with the different parts of the body.

GROSS MOTOR SKILLS

Gross motor skills refer to the ability to carry out activities that require large muscles or groups of muscles. Examples of gross motor tasks are walking, running, throwing something, jumping, standing on one leg, and swimming.

Fine Motor Skills

Fine motor skills consist of movements of small muscles that act in an organized and subtle fashion, for instance, the hands, feet, and muscles of the head (as in the tongue, lips, facial muscles), to accomplish more difficult and delicate tasks. Examples of fine motor activities are writing, sewing, drawing, pronouncing words (coordination of soft palate, tongue, lips), and whistling.

MOTOR PLANNING

Motor planning consists of the ability to imagine a mental strategy and than to carry out a movement or an action. Motor planning involves a number of abilities, including the visual detection of motion and errors in movement, selection of responses, and self-corrective motions. Movements must be timed adequately, and attention and concentration also are necessary.

Sequencing and Speed of Movements

Sequencing and speed of movements involves the order in which movements should proceed one after the other in order to accomplish a desired goal. It also is mostly unconscious or intuitive.

Test for Motor Sequencing

Touching the thumb against the other fingers of that hand in sequence.

Nose-finger Test

Performance in this tests measures fine motor coordination, proprioception, and perception of movements in space.

Moving a Limb Against Resistance (feet, legs, thighs, arms, forearms, hand)

This allows evaluation of strength in different areas.

Ability to Perceive Spatial Relationships

This ability depends on proprioception and the realization of where one's body is in space.

SENSORY INTEGRATION THERAPY

Sensory integration therapy is widely used in the treatment of children with certain difficulties that interfere with consistent, coordinated, effective motor function. Sensory integration therapy addresses the underlying

difficulties many clumsy children have with regulating, processing, and/or integrating sensory input. Accurate, coordinated, functional motor responses are based on well-modulated, selective, consistent information from both internal and external sources, which the brain then adequately integrates. Treatment involves specific input for the particular child and the facilitation of desired adaptive responses. Examples of internal senses are those of posture, proprioception, awareness of position of the body in general and of limbs and body parts in particular, i.e., vestibular input.

External stimuli that may facilitate or hinder performance may include auditory stimuli, lighting and visual stimulation, tactile sensations, and others. Children who are excessively impacted by such stimulation (who are therefore hypersensitive) are helped to cope with increasing amounts of stimulation, and the environment is modified to provide an acceptable level of sensory input. By contrast, in children who are hyposensitive, gradual increases in the amount of stimulation provided for the children are attempted. In all cases, signs of excessive or inappropriate stimulation are monitored.

Kinesthetic Training

Kinesthetic training relies on the studies on kinesthetic development in children by Laszlo and Bairstow. It deals with improving the kinesthetic sensitivity in children to improve motor control. The intent of kinesthetic training is to improve perceptual/motor dysfunction. It tries to generate improvement in the overall functioning of children in terms of perception and motion but does not focus on teaching specific skills as in other methods. Therefore, the theory behind this training is that when this general awareness of motion in space is improved, the motor skills also improve as a secondary effect.

Neurodevelopmental Treatment

The basic principles of treatment involve inhibition of primitive (persisting) reflexes and abnormal motor coordination patterns. On the other hand, it promotes the facilitation of higher-level reactions and normal muscular tone and movement patterns. Therefore, these authors emphasize creating a detailed evaluation of the patterns of development observed in the small child, making a diagnosis of the child's developmental level, and designing a strategy to restore development to as normal a level as possible and improve skills as much as possible.

Craniosacral Therapy

Osteopathic physician John E Upledger devised the technique of craniosacral therapy. Physiotherapists use highly sensitive palpatory skills that impact

imbalances in the craniosacral system. The treatment is reportedly successful mostly in those children who are most compromised in motor skills, who show some spasticity or hypertonia, and who have an abundance of health problems including motor coordination difficulties.

Visual Training

Visual training is an approach used to address specific difficulties with oculomotor function. Some visually related difficulties or difficulties with visual-motor integration interfere with coordinated motor skill performance as well as with equilibrium.

CHAPTER 37

Gait

NORMAL GAIT AND GAIT CYCLE

The body moves forward, one limb typically provides support while the other limb is advanced in preparation for its role as the support limb. A step is recognized as the interval between sequential floor contacts by ipsilateral and contralateral limbs. Two steps make up each gait cycle, which is roughly symmetric in normal individuals. The gait cycle is comprised of stance and swing phases.

Box 37.1 Summary of gait cycle (approximate duration is given in brackets)

The stance phase (60%) Initial double stance (10%) Single limb stance (10%) Terminal double limb stance (40%)
The swing phase (40%)

Gait Cycle Phasing

A consistent sequence of motions is performed at each of the lower extremity joints during locomotion. Each stride contains 8 relevant phases. Stance is comprised of 5 gait phases (i.e., initial contact, loading response, midstance, terminal stance, preswing), with the remaining 3 phases occurring during swing.

The first 2 gait phases (0–10% GC) occur during initial double support. These phases include initial contact and the loading response. Initial contact often is referred to as heel strike. The joint motion during this phase allows for transfer of weight onto the new stance phase leg while attenuating

shock, preserving gait velocity, and maintaining stability. Swing phase by the contralateral limb corresponds with single support by the ipsilateral limb to support body weight in the sagittal and coronal planes. The first half of single support is termed midstance (10–30% GC) and is involved with progression of the body center of mass over the support foot. This trend continues through terminal stance (30–50% GC). This phase includes heel rise of the support foot and terminates with contralateral foot contact.

The final stance element, preswing (50–60% GC), is related functionally more to the swing phase that follows than to the preceding stance phase events. Preswing begins with terminal double support and ends with toe-off of the ipsilateral limb.

ANALYSIS

Observational Gait Analysis

Appropriate to characterize most gait pathologies and sufficient to note gross abnormalities in walking.

Objective Gait Analysis

As walking complexity increases with organic pathology, objective analysis becomes necessary.

CLINICAL EXAMINATION

Routine assessment can be performed by a primary care physician; an expert may be needed for complex gait disorders. Assessment requires a straight hallway without distractions and a stopwatch for timing. A measuring tape and a T square or ruler with a right angle may be needed to measure stride length. Measurement of gait kinetics can only be performed reliably in a few laboratories with advanced computer and video technology.

Gait velocity is measured using a stopwatch. Step length (the distance from one heel strike to the next) can be measured or observed. Because shorter people take shorter steps and foot size is directly related to height, the easiest way to gauge step length is to measure or calculate the patient's foot length; normal step length is three foot lengths. Step height can be assessed by observing the swing foot; if it touches the floor, the patient may trip. Some patients with fear of falling or a cautious gait syndrome will purposefully slide their feet over the floor surface.

PATHOLOGICAL GAIT (TABLE 37.1)

Hemiparetic Gait

Hemiparetic gait characterizes spastic hemiparesis and is a common residual sign of a stroke. In this type of gait disorder shoulder is adducted and internally rotated, with flexion of the elbow, wrist, and fingers and with extension of the hip, knee, and ankle. Forward swing of the spastic leg during walking requires abduction and circumduction at the hip, often with contralateral tilt of the trunk.

Steppage Gait

Steppage gait is produced by weakness of ankle dorsiflexion. Because of the partial or complete foot drop, the leg must be lifted higher than usual to avoid catching the toe on the floor during the forward swing of the leg. Unilateral, steppage gait is usually due to L5 radiculopathy, sciatic neuropathy, or peroneal neuropathy. Bilateral, steppage gait is to a distal polyneuropathy or lumbosacral polyradiculopathy.

Paraparetic Gait

Paraparetic gait is a walking pattern in which both legs are moved in a slow, stiff manner with circumduction, similar to the leg movement as in hemiparetic gait. A paraparetic gait is due to the lesions of the spinal cord and also it is seen in patients with cerebral palsy. In many patients, the legs tend to cross with each forward swing ("scissoring").

Waddling Gait

In waddling gait due to weakness of hip flexion, the trunk is tilted away from the leg that is being moved to lift the hip and provide extra distance between the foot and the floor, and the pelvis is rotated forward to assist with forward motion of the leg. Because pelvic girdle weakness is customarily bilateral, the pelvic lift and rotation alternates from side to side, giving the waddling appearance to the gait. Waddling gait results from proximal lower limb weakness, most often from myopathy but occasionally from neuromuscular junction disease or a proximal symmetric spinal muscular atrophy.

Parkinsonian Gait

Parkinsonian gait is characterized by a forward stoop, with modest flexion at the hips and knees. The arms are flexed at the elbows and adducted at the shoulders, often with a 4- to 6-Hz resting pronation-supination tremor. Walking is initiated slowly by leaning forward and maintained with short rapid steps, during which the feet shuffle along the floor. The pace tends to accelerate (festinate) as the upper body gradually leans further ahead of the feet, whether movement is forwards (propulsion) or backwards (retropulsion).

Cerebellar Ataxic Gait

Cerebellar ataxic gait is a broad-based gait disorder in which the speed and length of stride varies irregularly from step to step. With midline cerebellar disease, posture is erect but the feet are separated; lower limb ataxia is also present in these patients.

Sensory Ataxic Gait

Impaired proprioception inhibits walking because of diminished information about the limb segment positions in space. Sensory ataxic gait may resemble a cerebellar gait, with its broad-based stance and difficulty with change in position. However, although balance may be maintained with the eyes open, loss of visual input through eye closure results in rapid loss of balance with a fall (positive Romberg sign).

Astasia-abasia (Hysterical gait)

Astasia-abasia is a typical hysterical gait disorder. Although the patient usually has normal coordination of leg movements in bed or while sitting, the patient is unable to stand or walk without assistance. If distracted, stationary balance is sometimes maintained and several steps are taken normally, before a dramatic demonstration of imbalance with a lunge toward the examiner's arms or a nearby bed.

Vestibular Gait

Vestibular gait is one in which the patient consistently tends to fall to one side, whether walking or standing.

Table 37.1 Important causes of specific gaits

Propulsive gait	Carbon monoxide poisoning Manganese poisoning Parkinson's disease Drugs including phenothiazines, haloperidol, thiothixene, loxapine, metoclopramide, and metyrosine (usually drug effects are temporary)
Scissors gait	Cerebrovascular accident (stroke) Cervical spondylosis with myelopathy Liver failure Multiple sclerosis Pernicious anaemia Spinal cord trauma Spinal cord tumour Syphilitic meningomyelitis Syringomyelia Cerebral palsy
Spastic gait	Brain abscess Brain tumour Cerebrovascular accident (stroke) Head trauma Multiple sclerosis
Steppage gait	Guillain-Barre syndrome Herniated lumbar disk Multiple sclerosis Peroneal muscle atrophy Peroneal nerve trauma Poliomyelitis Polyneuropathy Spinal cord trauma
Waddling gait	Congenital hip dysplasia Muscular dystrophy Spinal muscle atrophy

REHABILITATION ASPECTS

Ambulation always is associated with metabolic costs. These costs are relatively minor in normal adults performing free speed level walking. The self-selected walking speed in normal adults closely matches the velocity that minimizes metabolic work. This association does not apply with gait pathology. Walking velocity, energy cost per time, and energy cost per distance are considerations when the patient is making choices about walking versus wheelchair mobility. Gait velocity typically decreases with neuromuscular pathology, and the reduction is related to the magnitude of the pathology. Energy cost per unit of time is maintained by decreasing walking velocity considerably. Energy cost per unit of time does not change markedly following stroke, as compared to changes associated with aging; however, the energy requirement per distance traveled is more than 3 times normal. In this same population, wheelchair use cuts energy cost per distance in half and decreases cost per minute slightly, while preserving ambulation velocity. Similar trends are observed when examining various energy cost parameters in individuals with spinal cord injury, myelomeningocele, and increasing levels of amputation. Energy cost to travel a prescribed distance increases (greater than 500% increase in myelomeningocele with bilateral knee-ankle-foot orthoses), while oxygen cost per minute is maintained by decreasing walking velocity substantially. Often the critical factor in selecting a wheelchair for mobility is the energy requirement to traverse a given distance. Most individuals self-select wheelchair mobility when cost per distance exceeds 300% of normal values.

CHAPTER 38

Facial Paralysis

Facial nerve contains motor, sensory, and parasympathetic fibers vital for control of facial expression, taste to the anterior two thirds of the tongue, and salivary and lacrimal gland secretion. Of all the cranial nerves, the facial nerve is most susceptible to injury and facial paralysis can cause cosmetic inconvenience with associated functional problems.

CAUSES OF FACIAL PARALYSIS (TABLE 38.1)

Table 38.1 Causes of facial nerve palsy

Tumour	Schwannoma Neurofibroma Meningeoma Epidermoid Arachnoidal cyst Metastasis Paraganglioma Malignancy of the middle and external ear Parotid malignancy
Inflammatory	Multiple sclerosis Abscess Cerebritis Meningitis with facial nerve neuritis (bacterial) Facial neuritis with neurotopic virus (Bell's palsy and Ramsay Hunt Syndromes) Guillain-Barre´ syndrome Cholesteatoma Acute or chronic otitis media Parotid infection

contd.

Vascular	Stroke Arteriovenous malformation Haemangioma Subarachnoid haemorrhage Arterial aneurysm Vertebrobasilar aneurysm
Trauma	Temporal bone fracture (transverse and longitudinal) Post surgery Parotid gland biopsy
Miscellaneous	Osteopetrosis (Albers-Schonberg disease) Fibrous dyplasisa Paget's disease Diabetes mellitus Myasthenia gravis Hyperparathyroidism Mobius syndrome Sarcoidosis Melkersson-Rosenthal disease

CLINICAL EVALUATION

A thorough history includes onset, initial degree of paralysis, and associated symptoms. These details often can help identify the etiology (Table 38.2).

Table 38.2 Site of lesion and clinical features in facial nerve palsy

Site of the lesion	Clinical symptoms
Brain stem, cisternal segment, intracanalicular segment, labyrinthine segment, geniculate ganglion	Lacrimation, stapes reflex, taste and salivation are absent Hyperacusis Cranial nerve VI (brainstem lesion) may be affected Cranial nerve VIII (along the whole course) may be affected Motor branches of the occipital muscle, the posterior belly of the digastric muscle, stylohyoid muscle, mimic muscles of the face (in case of a brain stem lesion the temporal branches of the facial nerve are not affected), platysma are absent

contd.

Tympanic segment and mastoid segment up to the exit of the stapedius branch	Lacrimation is present Stapedius reflex, taste and salivation are absent Hyperacusis Motor branches of the occipital muscle, the posterior belly of the digastric muscle, stylohyoid muscle, mimic muscles of the face, platisma are absent
Between the exit of the stapedius and the chorda tympani branches	Lacrimation and stapes reflex are present Taste and salivation are absent Motor branches of the occipital muscle, the posterior belly of the digastric muscle, stylohyoid muscle, mimic muscles of the face, platisma are absent
Mastoid segment distal to the exit of the chorda tympani branch extracranial/intraparotid segment	Lacrimation, stapes reflex, taste and salivation are present Motor branches of the occipital muscle, the posterior belly of the digastric muscle, stylohyoid muscle, mimic muscles of the face, platisma are variably affected

Physical Examination

The physical examination of the patient with facial paralysis requires observation of the face at rest and during voluntary and reflex emotional movement. Determine total versus partial paralysis, unilateral versus bilateral involvement and degree of nerve dysfunction (Table 38.3). Assess symmetry at rest and during movement and the presence and degree of synkinesis. Note the severity of brow ptosis, ectropion, and oral commissure incompetence. Identify other cranial nerve or neurologic deficits and significant soft-tissue volume deficits in addition to the paralysis.

Table 38.3 Degree of nerve dysfunction

Grade	Degree of function
I	Normal
II	Slight weakness
III	Moderate weakness
IV	Moderately severe weakness
V	Severe weakness
VI	Total paralysis

INVESTIGATIONS

Electrophysiological

Electromyography with surface electrodes will confirm voluntary muscle contraction of the frontalis and platysma. An absent electromyogram from the lower eyelid and levator labii superioris whilst attempting to smile substantiated the clinical palsy.

Audiometry

Audiometric testing, including acoustic reflexes and tympanometry, may be useful in identifying the etiology of facial palsy secondary to retrocochlear pathology or mass lesions of the middle ear.

Radiography

High-resolution CT and MRI scans are essential in the evaluation of a patient with traumatic facial nerve palsy and in the patient with a possible tumour of the parotid, temporal bone, cerebellopontine angle (CPA), or skull base.

TREATMENT

Conservative

The ineffectiveness of traditional therapy methods for facial paralysis stems from their non-specificity and lack of adaptation to the unique characteristics of facial muscle. Facial muscle differs from most other skeletal muscle in several significant ways (Box 38.1).

Box 38.1 Facial muscle characteristics

- Lack muscle spindles
- Have small motor units
- Are relatively slow to degenerate
- Receive emotional as well as volitional neural inputs

Patients with traumatic facial paralysis often are treated empirically with a short course of oral steroids. In contrast to idiopathic facial paralysis or Bell palsy, no studies confirm or dispute the utility of steroid treatment following traumatic facial paralysis.

Proper eye care with artificial tears and night patching should be implemented as long as lid function is impaired.

Surgical Reconstruction

Both dynamic and static methods are used to reconstruct the paralyzed face. Dynamic procedures are far more successful in achieving the goals of reconstruction and should be considered in every patient presenting for facial reanimation.

Goals of Facial Nerve Reconstruction

Facial symmetry at rest
Adequate facial function, including oral competence and eye closure
Voluntary facial movement
Spontaneous facial expression
Absence of synkinesis or mass movement

Methods of Facial Rehabilitation

There are many procedures that have been described for surgical rehabilitation of the paralyzed face. These procedures can be grouped into those that restore neural input, those that replace nonfunctional facial muscles, those that statically resuspend facial tissues and adjunctive procedures that address specific defects.

Restoration of Neural Input

On lay nerve grafting
End-to-end anastomosis

- Without graft
- With graft:
  - Great auricular nerve
  - Sural nerve

Indirect Reconstruction

Cranial nerve crossover

- VII–XII anastomosis
- VII–XI anastomosis

Replace Nonfunctional Facial Muscles

Muscle transfer
Temporalis and masseter

Statically Resuspend Facial Tissues

Elevating the corner of the mouth or a ptotic brow (by using fascia lata or temporalis fascia)

Adjunctive Procedures

- Correct the lagopthalmos and ectropion
- Improve upper lid closure is the palpebral spring or gold weight
- Tarsorrhaphy

Summary

There are many possible treatment options for the patient desiring facial rehabilitation. No single procedure will address the cosmetic and functional deficits of every patient.Therefore, carefully tailoring the treatment plan to each individual's clinical picture will facilitate successful rehabilitation and lead to satisfied patients.

CHAPTER 39

Vestibular Diseases

Vertigo is defined as 'An illusion of movement' and is the hallmark of vestibular pathology.

MAINTENANCE OF NORMAL BALANCE

Balance is maintained by complex interactions within the CNS in which visual, vestibular and proprioceptive inputs are integrated. Vertigo will occur where there is a mismatch of sensory input, i.e., between vestibular and visual data.

CAUSES OF VERTIGO

Unilateral underactivity (paresis)
Trauma
Ischaemic damage
Viral labyrinthitis
Labyrinthectomy
Unilateral overactivity (irritative)
Acute (or chronic) ear infection
Meniere's disease
Benign paroxysmal positional vertigo

CLINICAL DETAILS

History

Associated symptoms
Hearing loss
Tinnitus
Ear infection – otorrhoea/otalgia
Visual loss

Musculoskeletal
Neurological e.g., sensory loss, headache

Examination

Otological to include hearing test
Neurological – cranial nerves, cerebellum, Romberg's test + Unterberger's stepping test
Ophthalmological – nystagmus, discs
Cardiovascular – carotids, arrythmia, carotid bruits
Musculoskeletal – where appropriate

INVESTIGATIONS

Audiograms
Vestibular function tests e.g., calorics, electronystagmography, sway platform
Radiological – MRI/CT scan

TREATMENT

Acute Labyrinthitis (Vestibular neuronitis/neuritis)

In the acute phase, vestibular sedatives (e.g., Stemetil, Stugeron) may be required
Bed rest.
In chronic or recurring cases
Central compensation can be improved by physiotherapy – vestibular rehabilitation exercises.

Benign Paroxysmal Positional Vertigo

In this condition, the patient experiences momentary vertigo on unilateral rotation of the head – often on turning over in bed – always to the same side, associated with nausea. It is common if there is a history of recent head injury or acute labyrinthitis.
Debris from the otolith organ gravitates into the most dependent portion of the posterior semicircular canal, such that head movements result in a prolonger movement within the canal, resulting in vertigo.
Head positional manoeuvres (Epley) are designed to reposition the debris, Brooksey-Cawthorne physiotherapy exercises may be helpful.

Meniere's Disease

Lifestyle advice – avoid overtiredness, stress, coffee, tea, red wine and cheese.
Medical - Serc (Betahistine), 8–16 mg, tds to reduce pressure, occasionally thiazide diuretics may help

Surgical
Endolymphatic sac decompression
Vestibular nerve section surgery for Meniere's disease is far from universally successful.

CHAPTER 40

Deafness

Hearing loss has a major contribution to communication and quality-of-life issues. Hearing loss can result in withdrawal, poor self-concept, depression, frustration, irritability, cognitive impairment, isolation, loneliness, and compromised physical mobility.

CAUSES OF HEARING LOSS

Conductive Hearing Loss

Conductive hearing loss results from dysfunction of the external or middle ear. There are four mechanisms, each resulting in impairment of the passage of sound vibrations to the inner ear.
Obstruction (e.g., cerumen impaction)
Mass loading (e.g., middle ear effusion)
Stiffness effect (e.g., otosclerosis)
Discontinuity (e.g., ossicular disruption)

Sensory Hearing Loss

Sensory hearing loss results from deterioration of the cochlea, usually due to loss of hair cells from the organ of Corti.
Advancing age (presbyacusis)
Excessive noise exposure
Head trauma
Systemic diseases (e.g., diabetes mellitus)

Neural Hearing Loss

Neural hearing loss occurs with lesions involving the eighth nerve, auditory nuclei, ascending tracts, or auditory cortex.
Acoustic neuroma

Multiple sclerosis
Cerebrovascular disease

EVALUATION OF HEARING (AUDIOLOGY)

Subjective Analysis

In a quiet room, the hearing level may be estimated by having the patient repeat aloud words presented in a soft whisper, a normal spoken voice, or a shout.

Tuning Forks

Tuning forks are useful in differentiating conductive from sensorineural losses. A 512-Hz tuning fork is employed, since frequencies below this level elicit a tactile response.

Weber Test

In the Weber test, the tuning fork is placed on the forehead or front teeth. In conductive losses, the sound appears louder in the poorer-hearing ear, whereas in sensorineural losses it radiates to the better side.

Rinne Test

In the Rinne test, the tuning fork is placed alternately on the mastoid bone and in front of the ear canal. In conductive losses, bone conduction exceeds air conduction; in sensorineural losses, the opposite is true.

Audiometric Studies

The site of the lesion responsible for sensorineural loss—whether it lies in the cochlea or in the central auditory system—may be determined with auditory brain stem-evoked responses.

MANAGEMENT OF AUDITORY IMPAIRMENT

Management of treatable conditions
Hearing aids
Speech (lip) reading
Use of hand signs, gestures
Writing boards

REHABILITATION IN CHILDREN

The most debilitating consequence of onset of hearing loss in childhood is its disruption to learning speech and language. The combination of early detection and early use of amplification will have a dramatically positive effect on the language acquisition abilities of a child with hearing loss. Aural habilitation/rehabilitation services for children typically involve:

Training in Auditory Perception

This includes activities to increase awareness of sound, identify sounds, tell the difference between sounds (sound discrimination), and attach meaning to sounds. Auditory perception also includes developing skills in hearing with hearing aids and assistive listening devices and how to handle noisy and difficult listening situations.

Using Visual Cues

It involves using all kinds of visual cues that give meaning to a message such as the speaker's facial expression, body language, and the context and environment in which the communication is taking place.

Improving Speech

This involves skill development in production of speech sounds (by themselves, in words, and in conversation), voice quality, speaking rate, breath control, loudness, and speech rhythms.

Developing Language

This involves developing language understanding (reception) and language usage (expression) according to developmental expectations.

Managing Communication

This involves the child's understanding the hearing loss, developing assertiveness skills to use in different listening situations, handling communication breakdowns, and modifying situations to make communication easier.

Managing Hearing Aids and Assistive Listening Devices

Because children are fitted with hearing aids at young ages, early care and adjustment is done by family members and/or caregivers. It is important for children to participate in hearing aid care and management as much as possible. As they grow and develop, the goal is for their own adjustment, learning, and troubleshooting of the hearing aid and, ultimately, taking on responsibility for making appointments with service providers.

REHABILITATION IN ADULTS

Learning to listen again
Assistive listening devices (hearing aid)
Using visual clues.

CHAPTER 41

Swallowing Dysfunctions

Dysphagia is a Greek word that means disordered eating. Dysphagia typically refers to difficulty in eating as a result of disruption in the swallowing process.

CAUSES OF DYSPHAGIA

Neurologic swallowing disorders are encountered more frequently in rehabilitation medicine than in most other medical specialties.

Neurologic Conditions

Stroke
Traumatic brain injury (TBI)
Motor neuron disease
Parkinson disease
Poliomyelitis
Multiple sclerosis
Myasthenia gravis
Myopathy (dermatomyositis, myotonic dystrophy)
Cerebral palsy and other movement disorders (mental retardation, developmental delay)

Non-neurologic Conditions

Laryngectomy
Pharyngectomy, esophagectomy reconstructed by gastric pull-up
Head and neck surgery (oral cavity cancer)
Cervical brace, cervical spondylosis
Ventilator-dependent patient
Elderly patients

isorders in the cervical esophageal aspect of deglutition
sophageal-pharyngeal backflow, tracheoesophageal [T-E] fistula, Zenker verticulum, reflux)

COMPLICATIONS OF DYSPHAGIA

Aspiration pneumonia
Malnutrition
Dehydration
Weight loss
Airway obstruction

MANAGEMENT

The goals of treatment are:
to maintain adequate nutritional intake
to maximize airway protection

Direct Strategies

Direct usually refers to treatment that involves food
Direct techniques include modifications of food consistency (Box 41.1)

Box 41.1 Different types of dysphagia diets

Thin liquids (e.g., fruit juice, coffee, tea)
Nectar-thick liquids (e.g., cream soup, tomato juice)
Honey-thick liquids (i.e., liquids are thickened to a honey consistency)
Pudding-thick liquids/foods (e.g., mashed bananas, cooked cereals, purees)
Mechanical soft foods (e.g., meat loaf, baked beans, casseroles)
Chewy foods (e.g., pizza, cheese, bagels)
Foods that fall apart (e.g., bread, rice, muffins)
Mixed textures

Thickened liquids increase oropharyngeal control. A diet of chopped or ced foods decreases difficulties with mastication.

Indirect Strategies

Indirect refers to an exercise regimen performed without food bolus
Direct techniques include stimulation of the oropharyngeal structures ption of behavioral techniques (such as postural changes or the swallow neuver)

Biofeedback

Biofeedback can be useful for oral motor and facial exercises. The patient

Enteral Feeding Methods (e.g., nasoenteric tube feeding)

Surgical Intervention

Surgery rarely is indicated in patients with oral or pharyngeal dysphagia, but it can be effective in selected patients.

- Surgical gastrostomy
- Cricopharyngeal myotomy

Compensatory Strategies

They are used to reduce the risk of aspiration and include the following:

Chin Tuck

The patient holds the chin down, increasing the epiglottic angles, and pushes anterior laryngeal wall backward, thereby decreasing the airway diameter.

Head Rotation

The ipsilateral pharynx is closed, forcing the food bolus to the contralateral pharynx while cricopharyngeal pressure is decreased.

Head Tilt

This technique guides the bolus to the ipsilateral pharynx using the effect of gravity.

Supraglottic Swallow

This technique involves simultaneous swallowing and breath-holding, closing the vocal cords and protecting the airway. The patient thereafter can cough to expel any residue in the laryngeal vestibule. The Valsalva maneuver may be used to maximize vocal cord closing.

Mendelsohn Maneuver

This maneuver is a form of supraglottic swallow in which the patient mimics the upward movement of the larynx by voluntarily holding the larynx at its maximum height to increase the duration of the cricopharyngeal opening.

CHAPTER 42

Spasticity

Spasticity is involuntary, velocity-dependent, increased muscle tone resulting in resistance to movement.

PATHOPHYSIOLOGY

Spasticity has many postulated causes, most revolving around altered afferent and efferent input to the alpha motor neuron. Spinal, peripheral nerve, or cortical injury can alter inhibitory and excitatory messages to the motor neuron. Alternatively, these injuries might result in denervation supersensitivity, deafferentation, central collateral sprouting, or disinhibition of the nerves. Polysynaptic responses may be involved in spinal cord-mediated spasticity, while enhanced excitability of monosynaptic pathways is involved in cortically medicated spasticity. Common causes of spasticity are listed in (Box 42.1).

Box 42.1 Common causes of spasticity

- Brain tumour/injury
- Cerebral palsy
- Multiple sclerosis
- Brain stem lesions
- Tethered spinal cord/injury
- Spinal cord tumour
- Hydrocephalus
- Intracranial, epidural, or subdural bleed

Advantages of Spasticity

- Substitutes for strength, allowing standing, walking, gripping
- May improve circulation and prevent deep venous thrombosis and oedema
- May reduce the risk of osteoporosis

Disadvantages of Spasticity

Orthopaedic deformity such as hip dislocation, contractures, or scoliosis
Impairment of activities of daily living (e.g., dressing, bathing, toileting)
Impairment of mobility (e.g., inability to walk, roll, sit)
Skin breakdown secondary to positioning difficulties and shearing pressure
Pain or abnormal sensory feedback
Poor weight gain secondary to high caloric expenditure
Sleep disturbance
Depression secondary to lack of functional independence

Measurement of Spasticity

Spasticity is difficult to quantify, but clinically useful scales include the following:
Ashworth scale – From 0–4 (normal to rigid tone) (Chapter 49)
Physician's rating scale – Assessment of gait pattern and range of motion
Spasm scale – From 0–4 (no spasms to >10/h)

CLINICAL ASPECTS

A lag time may exist between injury and spasticity onset, and severity may wax and wane over time. Spasticity may be static or dynamic in nature. Spasticity can wax and wane, appearing at variable times relative to date of injury or disease onset. Involved muscles may demonstrate spontaneous or elicited clonus, as well as increased deep tendon reflexes. Spasticity can occur in any muscle, but common patterns exist, especially when associated with an upper motor neuron injury (Table 42.1).

Table 42.1 Spasticity patterns

Pattern	Position of limb	Muscles affected
Upper extremity flexor patterns (CP, stroke, or TBI)	Adduction and internal rotation of the shoulder Flexion of the elbow and wrist Pronation of the forearm Flexion of the fingers and adduction of the thumb	Pectoralis major Latissimus dorsi Teres major Biceps Brachioradialis Brachialis Pronator teres and quadratus Flexor carpi radialis or ulnaris Flexor digitorum profundus and superficialis Adductor pollicis
Lower extremity patterns (CP, MS, stroke, and TBI)	Hip adduction and flexion Knee flexion Ankle plantar flexion or equinovarus positioning	Adductor magnus Iliopsoas Hamstrings (medial more often than lateral) Tibialis posterior Soleus Gastrocnemius
Extensor patterns (TBI)	Knee extension or flexion Equinus and/or valgus ankle Great toe dorsiflexion or excessive toe flexion	Quadriceps femoris Medial hamstrings Gastrocnemius Posterior tibialis Extensor hallucis longus Toe flexors Peroneus longus

condt.

Patient Evaluation

Examination

Detailed history and physical examination
Imaging studies of the head, neck, and spine or EMG or nerve conduction velocities (to rule out treatable causes)
To rule out any factors exacerbating spasticity (Box 42.2)

Box 42.2 Factors exacerbating spasticity

Infection (e.g., otitis, urinary tract, pneumonia) Pressure sore
Noxious stimulus (e.g., ingrown toenail, ill-fitting orthotics, occult fracture)
Deep venous thrombosis
Bladder distention
Bowel impaction
Cold weather
Fatigue
Seizure activity
Cold temperature
Malpositioning

Consultations

Plastic surgeons, orthopaedic surgeons, and neurosurgeons can play an important role in managing spasticity and its sequelae; thus, their contributions to the spasticity management team may be beneficial. Neurologists and urologists can assist with issues such as seizure control and neurogenic bladder, which may affect spasticity control.

Physical, occupational, speech, and recreational therapists can assist with family/patient training and education as well as therapeutic interventions.

TREATMENT

When deciding to treat a spastic muscle, it is important to assess the impact of its antagonistic muscle groups. Treatment of the agonist muscle without the antagonist may create an additional problem. For example spasticity may play a role in substituting for strength (i.e., to facilitate with transfers) in some patients.

Goals of Spasticity Management

To improve function with activities of daily living, mobility, ease of care for caregivers, sleep, cosmesis, and overall functional independence.
To prevent pressure areas from developing, orthopaedic deformity, and the need for corrective surgery.
To reduce pain
To allow stretch of shortened muscles, strengthening of antagonistic muscle, and appropriate orthotic fit

Box 42.3 Considerations that impact treatment

- Duration of spasticity
- Likely duration of therapy
- Severity of spasticity
- Location of spasticity
- Success of prior interventions
- Current functional status
- Future goals
- Underlying diagnosis
- Associated illnesses
- Patient compliance
- Availability resources

Treatment Approach to Patients with Spasticity

Preventative measures
Therapeutic interventions and physical modalities (Box 42.4)
Positioning/orthotics
Oral medications (Table 42.2)
Injectable medications
Surgical interventions
Prevention of alleviation or treatment of precipitating factors (Box 42.2)

Box 42.4 Physical modalities to treat spasticity

- Sustained stretching
- Massage
- Vibration
- Heat modalities
- Cryotherapy
- Functional electrical stimulation/biofeedback
- Strengthening of antagonistic muscle groups
- Hydrotherapy

Orthotics/Positioning to Treat Spasticity

Serial or inhibitive casting of the ankles, knees, fingers, wrists, and elbows
Splinting/orthotics: Upper and lower extremities, soft or hard, custom or prefabricated orthosis may help hold a limb in a functional position, reduce pain, and prevent deformity.
Positioning to reduce synergy patterns (e.g., wheelchair seating, bed positioning)
Children may require a new orthosis every few months due to growth. When newly casting, splinting, or positioning, monitor the skin closely for signs of breakdown.

Injectable Medications/Nerve Blocks

Phenol

Phenol is inexpensive, easily compounded, and has an immediate onset of action, usually in a 5% concentration.
Injected near motor points in the affected muscle.
Injections can be uncomfortable for some patients, and children may need to be sedated before injection.

Botulinum Toxin Type A or B

These medications are expensive but simple and fairly painless to inject. Injections of these substances block presynaptic release of acetylcholine at the neuromuscular junction. Collateral sprouting of the axon occurs in about 3 months, eliminating any permanent effect.

Combination Therapy

Botulinum toxin and phenol may be used effectively together to reduce the dose and improve the functional outcome.

Table 42.2 Common oral medications: Indications and side effects

Drug name	Mechanism of action	Site of action	Uses	Doses	Contraindications
Baclofen	GABA-B analogue, which presynaptically inhibits the nerve terminal	Centrally acting and can be administered intrathecally or orally	SCI and MS	Adult dose—5 mg PO tid; not to exceed 120 mg/d Paediatric dose10–60 mg/d PO	Documented hypersensitivity
Diazepam	Benzodiazepine, which acts presynaptically and is a GABA-A agonist	Centrally acting	SCI and MS	Adult dose—1 mg PO bid; titrate to effect; not to exceed 60 mg/d Paediatric dose 0.12–0.80 mg/kg/d PO	Documented hypersensitivity; narrow-angle glaucoma
Dantrolene	Prevents calcium release from sarcoplasmic reticulum	Peripherally acting medication	Effective in cerebral origin spasticity, such as in TBI, stroke, or CP	Adult dose—25 mg PO tid; titrate to effect; not to exceed 400 mg/d Paediatric dose—0.5 mg/kg PO bid to 3 mg/kg qid; not to exceed 400 mg/d	Documented hypersensitivity; active hepatic disease (hepatitis and cirrhosis)
Tizanidine	Muscle relaxant metabolized in liver and excreted in urine and feces	Centrally acting	SCI	Adult dose—2 mg PO tid; not to exceed 36 mg/d Paediatric dose—not established	Documented hypersensitivity
Clonidine	Stimulates alpha 2-adrenoreceptors in brain stem, activating an inhibitory neuron, which in turn results in reduced sympathetic outflow.	Centrally acting	Effective in SCI-associated and possibly TBI-associated spasticity.	Adult dose—1 mg PO bid; titrate to effect; not to exceed 2.4 mg/d Paediatric dose—5–30 mcg/kg/d PO	Contraindications Documented hypersensitivity

Surgical/Intrathecal Interventions

Tenotomy/Tendon Transfer/Osteotomy

Orthopaedic interventions release muscle contractures, lengthen shortened tendons, protect against or reduce bony deformities, and may reduce the strength of a spastic muscle group.

The timing of procedures is critical. If performed too early, repetitive procedures may be necessary or developmental milestones may be delayed. If delayed too long, future pain or irreversible bone deformity may occur. Orthopaedic interventions do not alter the spasticity of muscle groups inherently; they only alter the effects of spasticity.

Myelotomy/Cordectomy

Transection or resection of portions of the spinal cord result in reduced spasticity but potentially cause loss of bowel and bladder function, as well as loss of strength, pain, and temperature sensation.

These procedures rarely are performed.

Selective Dorsal Rhizotomy

Selective transection of the posterior spinal nerve roots from L2–S1 results in reduced lower extremity spasticity. Nerve roots are selected for ablation by evaluating the peripheral muscle and EMG activity that occurs during intraoperative stimulation.

Gait and upper extremity function as well as sensation also may be altered. This procedure appears to be most effective in a select group of young children with CP, who have strength underlying their spasticity.

Physical and occupational therapy are important postsurgical interventions to achieve the best outcome.

Spinal cord stimulator: Implanted percutaneously, stimulators currently are used more for pain reduction than reduction of spasticity, but they appear to be clinically effective in the future.

Stereotactic neurosurgery and cerebellar stimulation: These procedures have been used to reduce spasticity in some patient populations, but widespread benefits have not been noted.

Intrathecal Baclofen Therapy

Baclofen can be delivered intrathecally via an implantable pump placed under the skin or fascia of the abdomen. A catheter is attached and then tunneled into the intrathecal space, where it drips baclofen at a variable or continuous rate, 24 h/d.

The level of optimum catheter placement is determined clinically by the level of the muscle groups in which relaxation is desired.

Advantages
Small intrathecal dose is needed for effectiveness
Less sedation than oral baclofen
Effect lasts approximately 6–8 hours and predicts quantitatively, but not qualitatively, whether the medication will be effective. After the implant, the dosage of baclofen gradually is titrated until the desired effect is obtained.
The pump reservoir is refilled every 1–3 months via injection into a refill port.
The procedure is reversible.

Limitations
Catheter kinks or disconnection
Batteries may need replacement
Infections

CHAPTER 43

Pain Management

Pain is the most common symptom in medicine, usually a signal to the development of some potentially damaging lesion. Pain is often described in terms of a penetrating or tissue-destructive process (e.g., stabbing, pulling, twisting, tearing, squeezing) and/or of a bodily or emotional reaction (e.g., terrifying, nauseating, sickening).

PHYSIOLOGY OF PAIN

Pain may be classified into 'normal' pain and 'abnormal' pain.

Normal Pain

'Normal pain' is due to nociceptive stimuli such as scar, arachnoiditis, malignant infiltration or any such demonstrable lesion.

Abnormal Pain

'Abnormal pain' includes:

- ***Hyperalgesia*** (normal painful stimulus produce abnormally severe pain)
- ***Allodynia*** (gentle touch cause intensive pain)
- ***Hyperpathia*** (pain threshold is increased, but once reached it causes intense pain)
- ***Causalgia*** (above with features of sympathetic dystrophy such as shiny skin and tropic changes) are due to abnormal transmission

Pain Pathways

Previously, pain pathways were seen as having three components:

First order Neuron

First order neuron (cell body in dorsal root ganglion) which transmits from a peripheral receptor to the second-order neuron.

Second-order Neuron

A second-order neuron in the dorsal horn of the spinal cord, which axon crosses the midline to ascend in the spinothalamic tract to the thalamus where it synapses with third-order neuron

Third-order Neuron

A third-order neuron projects to the postcentral gyrus (via the internal capsule).

Recently for better understanding of pain pathways it has been divided into the following components;

- Peripheral receptors
- Neural pathways
- Spinal cord mechanisms and long tracts
- Brainstem, thalamus, cortex and other areas.
- Descending pathways.

a) Peripheral Receptors

In peripheral receptors there is the conversion of one form of energy, (thermal, mechanical, or chemical), into a form that is accessible to the brain (nerve impulse). A painful stimulus causes a "bright," sharp, localized sensation followed by a dull, intense, diffuse, and unpleasant feeling. These two sensations are variously called fast and slow pain or first and second pain. Fast pain is due to activity in the A delta pain fibers whereas slow pain is due to activity in the C pain fibers.

First pain (fast pain) is described as sharp, and "pricking". It localises to a well-defined part of the body surface. The receptors for this first pain are high threshold mechanoreceptors.

Second pain (slow pain) is due to stimulation of receptors that exist in many tissues (but not in, paradoxically, the brain). It is often described as dull (i.e., not sharp) and aching. It is poorly localized. Visceral pain is predominantly of the "second pain" type. Visceral pain can however sometimes be referred to a region of the body surface (for example, shoulder tip pain with subdiaphragmatic irritation).

Neurotransmitters

There is some evidence that neurotransmitters such as substance P, vasoactive intestinal polypeptide (VIP) and calcitonin gene-related peptide are important mediators, either as neurotransmitters, or sensitizers of visceral pain receptors. Prostaglandins, histamine, serotonin, bradykinin, ATP, potassium, and H^+ ions also appear important in this regard, especially serotonin, which appears to act mainly on 5HT3 receptors.

...n Perception

...terms of pain perception, thresholds for feeling pain are remarkably ...istant from individual to individual i.e., peripheral receptor stimulation ...sufficient intensity will reproducibly cause pain at the same level in ...st people. The response of the individual, and his tolerance of the pain, ...ll however differ markedly between individuals (Box 42.1).

Box 43.1 Factors modifying perception of pain

Circumstances in which the injury is sustained
Anticipation of pain
Past experiences of pain
Psychological factors (i.e., mood and state of distraction)

...o Neural Pathways

...st pain" responses are conveyed from the periphery to the dorsal horn ...the spinal cord in ***small myelinated fibres*** (A delta) while "second" ...n is conveyed in ***non-myelinated C fibres***. (Table 43.1)

Table 43.1 Properties of A-delta and C-fibers

A-Delta	C-Fiber
Myelinated, large	Non-myelinated, small
Respond to mechanical stimuli and some to thermal also	Responds to any noxious stimuli.
Receptive fields consist of cluster of small spots	Receptive field is a single area rather than clusters
May be sensitized	May be sensitized.
Resistant to local anaesthetics but susceptible to pressure	Susceptible to local anaesthetics
Inactivated with higher temp	Inactivated at temp 55°C
Responsible for I pain (early, sharp, brief pain)	II pain (dull, prolonged pain)

c) Spinal Cord Pathways

Spinal cord grey matter is organized into ten laminae (REX) C-fibers project mainly to I and II laminae and A-delta to I to V. The neurons in the cord can be divided into projection neurons (relay to higher centers), excitatory neurons (relay to projection and other interneurons or to motor neuron that mediate spinal reflexes) and inhibitory interneurons (contribute to the control of nociceptive transmission). Laminae I, V, VII, VIII are the major sources of rostarlly projecting nociceptor neuron. Laminae II (substantia gelatinosa) make predominantly local connection that result in important changes in the neuronal activity. Unmyelinated C fibres synapse in laminae I to V, while A-delta fibres synapse in laminae I, V and X. The major spinal pathways for pain travels in the anterolateral spinal quadrant to the thalamus (spinothalamic tract) and crosses over to the other side 2 or 3 segments above. Certain proportion of pain impulses can be carried in ipsilateral pathway.

d) Higher Ascending Pathways

Spino-reticulo-diencephalic Pathway

Spino-reticulo-diencephalic pathway (old) mainly ends in the reticular system of the brainstem, also sends fibres to the thalamus (the medial nuclei of the thalamus). Probably important are connections between the reticular system and the hypothalamus (and thalamus)—these may explain autonomic components of the pain response.

Spinothalamic Tract

Spinothalamic tract (new) nips up to the ventrobasal part of the lateral thalamus. Connections go from here to the sensory cortex (postcentral gyrus), which explains the precise localisation of somatic pain.

The Spinothalamic Tract Divides Into

- Lateral division which terminates in posterior nuclear group and ventrobasal nuclei (VPM & VPC). The major projection is from I & V laminae with receptive field restricted to one side of the body usually part of a limb.
- Medial division is called paleospinothalamic tract and terminate in the central lateral nucleus. The major projection is from entire body surface.
- Spinoreticular projection appears to involve V, VI VII & VIII laminae and have complex receptive fields from both sides similar to paleospinothalamic tract.

e) Descending Pathways

Descending modulation of pain sensation originates from three main areas:

- Cortex
- Thalamus
- Brainstem, where the *periaqueductal grey matter* (PAG) is particularly important.

THE RESPONSE TO PAIN

Responses to visceral pain are very different from those evoked by somatic pain. Visceral pain generally results in tonic muscular spasm (to decrease movement of the affected area) while somatic pain usually causes withdrawal of the affected part of the body ("to protect this region from further damage").

PAIN MODULATION

The process by which the nervous system modifies the nociceptor activity is called modulation. The modulatory network is quite different from sensory system and involves a number of brainstem regions (periaquenductal grey and immediately adjacent midbrain, periventricular grey of hypothalamus, the lateral and dorsolateral pontine tegmentum and rostroventral medulla). Stimulation of any of these sites reduce pains and inhibit nociceptive neurones. Both nor-adrenergic and serotonerigc systems are involved.

NEUROPATHIC PAIN

The term neuropathic pain includes all types of pain due to lesions of the sensory pathway, both peripheral and central. Conditions affecting the primary sensory neurone (including the dorsal root ganglion and dorsal root) are much more common than the central causes. The many causes of neuropathic pain are shown in Table 43.2.

Clinical Features of Neuropathic Pain (Box 43.2)

The onset of neuropathic pain may be immediate following the initiating disease, but is often delayed. Changes in emotional state and fatigue may cause the pain intensity to vary over short periods.

Spinal Cord Lesions

Pain from spinal cord lesions may be localized, unilateral, or bilateral, but often diffuse and widespread below the level of the lesion. The pain is usually continuous and may have an aching, stinging, burning, cramping, or girdle-like quality. Both the ongoing and paroxysmal pains are usually provoked but either may be exacerbated by movement, fatigue, or emotion.

Table 43.2 Causes of neuropathic pain

Peripheral nerve	Trauma Entrapment neuropathies Partial or complete transection Painful scars Mononeuropathies and multiple mononeuropathies Diabetes Malignant nerve/plexus invasion Radiation plexopathy Polyneuropathies (alcoholic, nutritional, Isoniazid)
Root and dorsal root ganglion	Prolapsed disc Post-herpetic neuralgia Arachnoiditis Trigeminal neuralgia Root avulsion Tumour compression Surgical rhizotomy
Spinal cord	Complete transection Tethered cord Vitamin B_{12} deficiency Trauma Syringomyelia Multiple sclerosis Tumours Arteriovenous malformation
Brain-stem	Wallenberg's syndrome Multiple sclerosis Syringobulbia Tuberculoma Tumour
Thalamus	Infarction Haemorrhage Tumours
Cortical and subcortical	Infarction Tumour Arteriovenous malformation Trauma

Box 43.2 Clinical characteristics of neuropathic pain

Quality of pain: often burning, raw, aching, gnawing
Additional paroxysmal, shock-likepains
Pain associated with some sensory impairment
Associated allodynia, hyperalgesia, and hyperpathia are common
Sympathetic dysfunction is common, particularly with peripheral nerve lesions
Sometimes associated with changes of reflex sympathetic dystrophy
The onset of pain may be immediate or delayed
Pain intensity is often markedly altered by fatigue and emotion

Brain-stem Lesions

Vascular lesions affecting the pons and medulla are the commonest brain-stem lesions leading to pain. Multiple sclerosis, tumours, syrinx, and tuberculoma are occasional causes. Lesions confined to the midbrain do not cause pain.

Thalamic Lesions

Pain was originally described as part of the thalamic syndrome by Dejerrine and Roussy in 1906. The syndrome comprised superficial and deep hemianaesthesia, sensory ataxia, intractable pain, mild hemiplegia, and sometimes choreo-athetosis. It is almost always caused by infarction, with haemorrhage and arteviovenous malformation being occasional causes and, very rarely, tumours involving the thalamus.

Cortical and Subcortical Lesions

Lesions rostral to the thalamus leading to pain are extremely rare but vascular lesions, trauma, and tumours are recorded causes. Pain has also been observed after therapeutic parietal cortectomy for intractable pain (not an operation now performed) and after hemispherectomy for severe epilepsy with infantile hemiplegia, or for the treatment of tumour.

MANAGEMENT

Acute pain is an alerting or useful pain, signifying tissue injury from a medically or operatively remediable somatic cause, often accompanied by signs of autonomic hyperactivity. Chronic pain, however persists beyond the period of what was believed to be curative treatment. Multidisciplinary pain clinics consist of a core staff that includes an anaesthetist, physiotherapist, occupational therapist, and psychologist, but the availability of other specialists, particularly a neurologist and a psychiatrist, is important.

Local Measures

Most pains are localized and local measures should always be attempted in the first instance, although a combination of local and systemic treatment may be necessary.

Counter Stimulation

Regardless of the type of pain, whether nociceptive or neuropathic, counterstimulation techniques may be helpful.

Heat and Cold

Many intractable musculoskeletal pains respond to heat, which can be delivered by a heat pad or radiant heat. Neuropathic pains tend to be exacerbated by cold but an exception is post-herpetic neuralgia in which regular application of cold packs for 20 min may reduce troublesome allodynia for up to several hours.

Vibration and Ultrasound

Both nociceptive and neuropathic pains may respond to these simple measures, of which vibration is the more practical on a long-term basis since it can be used regularly by the patient at home.

Transcutaneous Electrical Stimulation (TENS)

TENS, acting by segmental inhibition, has been shown to be effective in a wide range of different chronic pains, but is probably more effective for neuropathic than nociceptive pains. TENS may produce analgesia over a wider area than can be accounted for on the basis of segmental inhibition, and this is probably due to the diffuse noxious inhibitory control mechanism already discussed. A trial period of 2 to 3 weeks, with the patient experimenting with different electrode placements, is essential before concluding that TENS is unhelpful.

Acupuncture

Acupuncture probably works by segmental inhibition and by diffuse noxious inhibitory control. The latter is more marked with the noxious counterstimulation of acupuncture than with TENS. Cerebrospinal fluid endorphin levels are raised by acupuncture but not by TENS, and TENS-induced analgesia is not reversed by naloxone.

Topical Local Anaesthetic

Simple lignocaine ointment, although poorly absorbed, is sometimes helpful in areas of severe allodynia and hyperpathia, particularly in some patients with painful scar syndromes, tender amputation stumps, and post-herpetic neuralgia.

Topical Capsaicin

Capsaicin (0.075 per cent) may also be effective in these situations, probably through its action of desensitizing afferent C fibres for long periods. However, it may produce intolerable burning pain in some patients.

Psychological Techniques

Psychological techniques such as hypnotherapy, meditation, stress management techniques, relaxation training or biofeedback will help.

REHABILITATION

Rehabilitation should be included in all treatment plans. Occupational therapy and vocational rehabilitation may also be appropriate.

Drug Therapy

Simple analgesics (such as paracetamol and aspirin) the NSAIDs, weak opiates (including codeine, dihydrocodeine, and dextropropoxyphene), partial opiate agonists (such as buprenorphine) mixed agonist antagonist drugs (for example pentazocine), and strong opiates may have a beneficial effect in nociceptive pains. Analgesics should be prescribed as round the clock medication to be effective and decrease the total drug required. Simpler drugs (aspirin) should be maximized before switching to stronger alternatives. Adjunctive drugs include those specific for the etiology (e.g., phenytoin, carbamazepine of trigeminal neuralgia, NSAID or muscle relaxants for chronic soft tissue and muscle changes). Pain modulation may be stimulated by tricyclic antidepressants (amitriptyline or doxepins).

Local Anaesthetic Injections

Local anaesthetic blocks to peripheral tissues, peripheral nerves, or roots may be useful in following ways.

- First, the origin of a particular pain may be more accurately defined.
- Secondly, injection into a peripheral trigger point, for example in muscle, may reduce widely radiating pain and offer effective treatment with one or a series of injections.
- Thirdly, in the case of neuropathic pain due to peripheral nerve or root lesions, failure to relieve all the pain with an adequate peripheral nerve or root block indicates an element of secondary central pain, which will be less amenable to peripheral measures.

Lesions including trigger points in muscle, painful scars, and peripheral nerve lesions such as neuromas may respond better to a combination of local anaesthetic and corticosteroid. Epidural injection of a local anaesthetic, with or without an opiate, can produce analgesia over a wide area.

NEUROSURGICAL INTERVENTION

Neurosurgical procedures can be divided into stimulation procedures or ablative procedures and briefly mentioned below.

Stimulation Procedures

Stimulation procedures depend on blocking pain pathways or reversible stimulation of inhibitory pathways and do not ordinarily result in destruction and hence are reversible.

Transcutaneous Stimulation

Transcutaneous stimulation, in which a controlled electrical stimulus to the skin is a popular one with about 50% success rate regardless of cause of pain. According to gate theory when large myelinated fiber activity is increased by non-painful stimulus, the pathway for non-myelinated small fibers transmitting pain is closed. Rubbing an injured part similarly reduces pain.

Peripheral Nerve Stimulation

Peripheral nerve stimulation is similar to the above with electrode around the individual peripheral nerve in neuropathic pains confined to a single nerve.

Dorsal Column Stimulation

Dorsal column stimulation applies a train of electrical stimuli to the dorsal aspect of the cord by means of an apparatus that can be controlled by the patient. This attempts to stimulate the collaterals of the large fibers as they ascend in the dorsal column, resulting in increasing the rate and inhibits perception of pain.

Deep Brain Stimulation

Deep brain stimulation is an outgrowth of the above and still is an investigational procedure, despite the development of steretactic procedures. The internal capsule, ventral posterior nuclear complex are the usual targets. This helps in pain secondary to cord lesions, thalamic syndrome, or phantom limb pains. Periventricular or periaqueductal grey is a recent target and related to endogenous opiate analgesia.

Destructive or Ablative Procedures

Destructive lesions or ablative procedures deprive the patient of pain and possibly of other sensations and are not reversible.

Peripheral Nerve Blockade

It is of limited value, but easily available, also serve to test the possible result of permanent denervation.

Posterior Spinal Root Blockade

Judicious amount of phenol or ethyl alcohol into the spinal subarachnoid space would damage the adjacent sensory rootlets sufficiently to block afferent impulses for several months. Subarachnoid injections are most effective from the low thoracic level. At higher levels, some prefer extradural injections. Depending on the concentration and duration of exposure, phenols could cause reversible or irreversible block. Immediate effect is that of a local anaesthetic. The permanent effects are due to degeneration. Long acting steroid (Depomedrol) is often used these days along with local anaesthetics especially in chronic pain of radicular origin.

Posterior Rhizotomy

If the course of pain can be accurately delineated by segmented boundaries and is limited to few divisions, rhizotomy should provide permanent relief. Such conditions include traumatic lesions of peripheral nerves, operations scars, intercostal or occipital neuralgias.

Anterolateral Cordotomy

Extremely useful in the management of chronic pain.

Open Cordotomy

Open cordotomy is usually performed at one of the two levels, the T3 for pain below midthoracic level and C1–2 for pain above the midthoracic level.

Percutaneous Cordotomy

Percutaneous cordotomy requires a cooperative patient and specialized equipments and is recommended for unilateral pain. It involves physiological localization in an awake patient and graded radio frequency electrical destruction of the tract.

Dorsal Root Entry Zone (DREZ) Cordotomy

A destructive lesion is created in the posterolateral sulcus of the spinal cord at the point of entry of the dorsal roots. DREZ lesions are designed to destroy regions of neuronal dysfunction in deafferentation states involving particularly Lissauer's tract and I,II & V Rex layers. DREZ lesions help in deafferentation pain such as causalgia, root avulsions, herpetic neuralgia etc.

Commisural Myelotomy

The spinothalamic fibers can be interrupted as they cross the anterior commissure by a vertical incision in the median plane. The result is bilaterally symmetrical area of analgesia. This procedure is not widely accepted.

Spinothalamic Tractomy in the Brainstem

Medullary, pontine, mesencephalic tractomies have been described. Due to high mortality and morbidity, they never became popular. Stereotactic techniques have also been tried. It may have a role in cancer pain involving the head and neck.

Sympathectomy

Repeated temporary anaesthetic blockade should proceed symphathectomy. Main indications for symapthectomy are causalgia, sympathetic dystrophy and painful ischaemic states.

Stereotaxic Thalamic Lesioning

This involves lesioning thalamic nuclei and hence disturbs nociception. There is a shift of stereotaxic lesions away from ventral lateral (specific) nuclei to the ventral posterior medial and interlaminar (nonspecific) nuclei. Since the introduction percutaneous cordotomy, the thalamotomies have became rare.

Operations on the Cortex and Subcortex

The aim is to create lesions deep to secondary sensory which severs its links with thalamus. Leucotomy was once practiced. Stereotactic cingulomotomy and inferomedial quadrant frontal section have proved helpful. All these aim to disturb pain perception.

CHAPTER 44

Nutritional Aspects

After the brief, metabolically quiescent period (24–48 h) that may follow the initial insult, a hypercatabolic state develops, such that, in the absence of an exogenous supply of protein, energy, vitamins and trace elements, a rapid loss of muscle protein occurs and the patient becomes progressively more susceptible to the complications of malnutrition.

INDICATIONS

Virtually all patients appropriately admitted to intensive care for more than 3 days.

Severe injury, surgery, and sepsis comprise the majority of admissions where prompt nutritional support is necessary to limit the extent of the negative nitrogen balance.

ASSESSMENT

It is important to assess the baseline nutritional and fluid balance status on admission and to review both fluid and calorie requirements daily.

Thorough medical and nutritional history

Details of recent weight change and calorie and fluid intake

Extent of any weight loss and muscle wasting

Evidence for specific nutrient deficiencies

Precise monitoring of body weight (extremely difficult in intensive care)

Input/output charting

BLOOD TESTS

Serum albumin

Transferrin, prealbumin, and retinol-binding protein are more sensitive tests but generally less readily available.

Total nitrogen losses may be determined from direct measurement of urinary nitrogen excretion and estimation of other losses, particularly from the gastrointestinal tract.
Energy expenditure may be calculated by indirect calorimetry from measurements of carbon dioxide production and oxygen consumption.

PARENTERAL NUTRITION

Depends on the diagnosis.
Total parenteral nutrition will be required in approximately 10 per cent of patients.
Requires suitable central access and safe prescription, preparation, and delivery.
Provides the advantage of simultaneous delivery of the nutrients.

Indications

Patients with a nonfunctioning GI tract
Patients unable to tolerate oral or enteral nutrition

Routes

Peripheral parenteral nutrition (PPN)
Central or total parenteral nutrition (TPN)

Complications

Paralysis
Glucose intolerance
Anaemia
Paralytic ileus
Gastrointestinal ulcers
Neurogenic bowel and bladder
Depression
Skin/wound breakdown
Pneumonia

ENTERAL FEEDING

Provided there is no absolute surgical contraindications or significant risk of lung aspiration, enteral feeding is always preferable to parenteral nutrition.

Advantages

Safe
More physiological

Easy to administer
Requires less medical and nursing expertise
Avoids both the risks and cost of parenteral nutrition
Helps to maintain gut mucosal integrity

Enteral Feeding Routes

Nasogastric
Nasoduodenal
Gastrostomy
Jejunostomy

Complications of Enteral Feeding

Complications associated with enteral support can be avoided by proper formula selection, proper administration, and careful monitoring.
Diarrhea
Malabsorption
Bacterial contamination
Lactose intolerance
Lack of fiber
Altered bacterial flora
Nausea/vomiting
Aspiration

NUTRITIONAL REHABILITATION

The goal of nutritional rehabilitation is to optimize intake while the patient is hospitalized to promote wound healing and to provide energy to undergo therapy. One should remember that optimal nutrition cannot be achieved if the patient is not inspired to eat the foods provided (Box 44.1).

Box 44.1 Principles of nutritional rehabilitation

Nutrition is the one treatment modality over which the patient usually has control
Avoid complicated diet prescriptions
Maintain a liberal attitude, providing the diet that does not endanger patient's condition
Adjust the patient's diet according to need
If intake does not improve with dietary modification, than prescribe supplementation.
If the patient has any upper extremity disabilities, independent feeding may be hampered
Find out conditions that may affect eating (ill-fitting dentures, missing teeth, edentulous poor dental hygiene, oral infections)
Discuuss the diet with the patient

Table 44.1 Management options

Gastric irritation	Avoid dry/salty foods Offer moist/soft foods
Nausea/vomiting	Small quantities of foods that are easily digestible Maintaining adequate hydration Avoiding fried/greasy foods Cold instead of hot food
Eating environment	Clean healthy surroundings
Lighting	Proper lighting Let the patient see what he/she is going to eat.
Seating	Transfer the patient to a chair for meals if possible Adjust the tray table to proper height
Positioning	Proper positioning improves the comfort level
Room occupancy	The ideal environment—discuss with the patient Encourage family members to visit at mealtime
Follow up	Direct observation Nutritional assessment

Orthotics

INTRODUCTION

Orthosis (or orthotic device) is the medical term that refers to a brace or splint. Orthoses are devices applied externally to restore or improve functional and structural characteristics of the musculoskeletal and nervous systems.

TERMINOLOGY

Orthotics

Orthotics is defined as the unit of rehabilitation which deals with improving function of the body by the external application of a device which aids the body part.

Orthosis

An external device applied on the body to limit motion, correct deformity, reduce axial loading or improve function in a certain segment of the body.

Caliper

Caliper is a specialized type of orthosis prescribed for lower limbs and the main component material is metal.

Splint

Splint is a device used for immobilization of part of a body.

TYPES OF ORTHOSES

Static Orthoses

As the word static implies, these devices do not allow motion. They serve to provide a rigid support in fractures, inflammatory conditions of tendons and soft tissue, and nerve injuries.

Dynamic/Functional Orthoses

In contrast to static orthoses, these devices do permit motion on which its own effectiveness depends. These types of orthoses are used primarily to assist movement of weak muscles.

CLASSIFICATION OF ORTHOSES

One single classification is difficult and hence GK Rose grouped them as follows:

1. Functional biomechanical orthoses
2. Functional descriptive orthoses
3. Nosological (according to disease) orthoses
4. Regional orthoses

FUNCTIONS OF ORTHOSES

- Stabilizes flail joints
- Prevents deformities
- Corrects deformities
- Prevents unwanted movements
- Facilitates useful movements
- Provides rest to a part.

TRAINING OF PATIENT TO USE AN ORTHOSES

- If patient receives an orthoses, he should understand the function of orthoses
- How to put and take off/look after it
- To enable him to acquire the maximum benefit from its use
- Excercises to strengthen certain groups of muscles
- Walking aids like parallel bars may be necessary initially
- Once the patient can walk with confidence must be taught to walk up and down inclines, and to climb up/down steps.
- How to sit and lie on the floor, pick up objects from floor is instructed.

CARE OF THE ORTHOSES

- Handle with care and avoid dropping it
- Examine skin every night for evidence of undue pressure from orthoses
- Open all locks and remove any dust or fluff weekly
- Oil each joint weekly
- Inspect all moving parts of wear, bolts, screws for loosening tight
- Inspect all leather parts regularly, get any necessary repair
- Keep heels and soles of the footwear in good condition.

MATERIALS USED IN ORTHOSIS

An orthosis can be constructed from metal, plastic, leather, synthetic fabrics, or any combination. Plastic materials, such as thermosetting and thermoplastics, are the materials most commonly used in the orthotic industry (Table 45.1).

Table 45.1 Materials used in orthotics

Material	Advantage	Limitations	Uses
Plastic	Molded into permanent shape after heating	Do not return to their original consistency	Low-temperature thermoplastics—used mainly in low stress activities High-temperature (polypropylene) thermoplastics—used for high stress activities
Metal (stainless steel aluminum alloys)	Adjustable	Heavy Cosmetically unpleasant	Joint components Metal uprights Sprints Bearings
Leather (i.e., cattle hide)	Conducts heat Absorbs water		Shoe construction
Rubber	Tough resiliency Shock-absorbing qualities		Padding in body jackets and limb orthoses

Box 45.1 Drawbacks of orthotic device use

Discomfort
Local pain
Osteopenia
Skin breakdown
Nerve compression
Muscle atrophy with prolonged use Increased energy expenditure with ambulation
Difficulty donning and doffing orthosis
Difficulty with transfers
Increased segmental motion at ends of the orthosis

UPPER EXTREMITY ORTHOSES

Functions of Upper Extremity Orthoses

To increase range of motion (ROM)
To immobilize an extremity and to help promote tissue healing
To apply traction either to correct or prevent contractures
To assist in providing enhanced function
To serve as an attachment for assistive devices
To help correct deformities
To block unwanted movement of a joint

Table 45.2 Arm orthoses

Type	Indications	Examples
Arm sling	Scapular or humeral fractures Acromioclavicular joint injury Rotator cuff injury Bicipital tendinitis Hemiparesis with subluxation	Figure-8 sling Cuff sling Glenohumeral support
Functional orthoses arm	Proximal arm weakness involving the shoulder and arm (i.e., SCI and peripheral nerve lesions)	Shoulder saddle suspending a proximal forearm cuff by straps or Bowden cable
Balanced forearm orthosis	High-level tetraplegia or severe proximal arm weakness or paralysis Supports the weight of the forearm and arm against gravity	Patients may be able to perform tabletop activities.
Posterior elbow splints	Elbow immobilization (elbow surgery and or inflammation)	
Serial cast	Prevention or correction of contractures by promoting soft tissue stretch and passive ROM	
Air splint	To maintain or increase elbow extension (contractures and elbow immobilization)	Circumferential inflatable sleeve
Dynamic elbow flexion orthosis	Used to maintain the elbow in 90° of flexion	Elbow contractures, burns and fractures

Arm Sling Pouch (Figure 45.1)

It is a simplest and most common orthotic devise used to position the arm. The presence of non-stretching strap helps to reduce pressure and evenly distribute weight of affected arm over the other shoulder.

Indications

Support the arm or shoulder in case of closed or surgical management of:

- Strain or sprain
- Fractures and dislocations – either as an emergency management or even for post-operative splinting.

Correct method of application:

Flexion of the elbow greater than 90°, as it aids in limb elevation and thereby decreases dependent oedema. Wrist should also be supported in the sling.

Advantages

- Low cost
- Ease of use
- Light weight and
- Portability

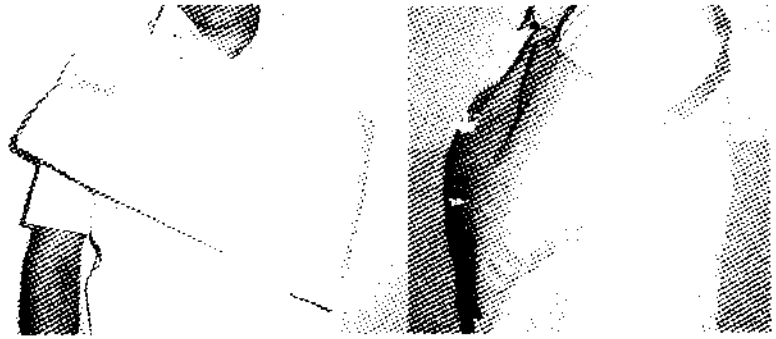


Fig. 45.1 Arm sling pouch

Clavicular Brace (Figure 45.2)

It is made of padded straps that form a 'figure of eight' compression over the clavicular region. The shoulder on the affected side must be pushed up during application of the clavicular support.

Indications

- Management of clavicular fractures
- Acromio-clavicular and scapular separations
- Posture correction in case of stooped shoulders

Fig. 45.2 Clavicular brace

Free Flexion Extension Brace (Figure 45.3)

It is designed to permit free range of flexion and extension without increased lateral or anteroposterior mobility.

Indications

- Any condition resulting in painless hypermobility such as joint resection, non union of bones of arm or forearm.

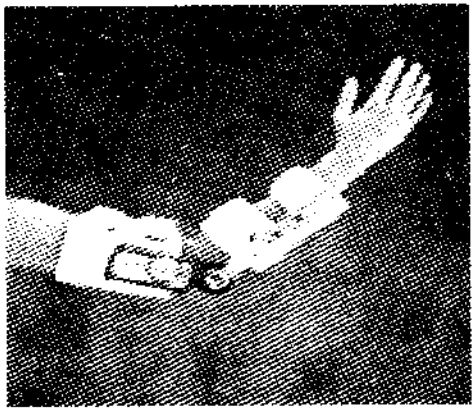


Fig. 45.3 Free flexion extension brace

Turnbuckle Orthosis (Figure 45.4)

It aims to increase elbow range of flexion and extension.

Indications

- Soft tissue contractures (post-traumatic, post infectios, neuromuscular)
- Type of static progressive splint.

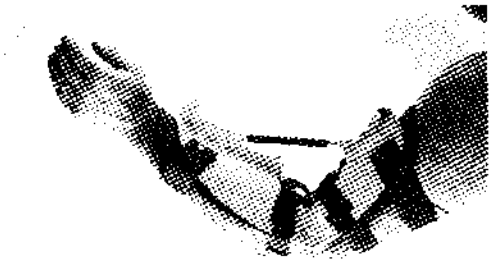


Fig. 45.4 Turnbuckle orthosis

Cock-up Splint (Figure 45.5)

These are devices designed to maintain the position of dorsiflexion of the wrist or to force the joint into that position.

Two types available:

- Static
- Dynamic

Indications

- Sprained wrist
- Wrist drop
- Minor fractures of the wrist
- Following early cast removal

Fig. 45.5 Cock-up splint

Carpal Tunnel Splint (Figure 45.6)

It is a splint to keep the wrist joint in neutral position, so as to prevent median nerve from compression in positions of dorsi and palmar flexion.

Indications

- Management and prevention of recurrence of carpal tunnel syndrome.

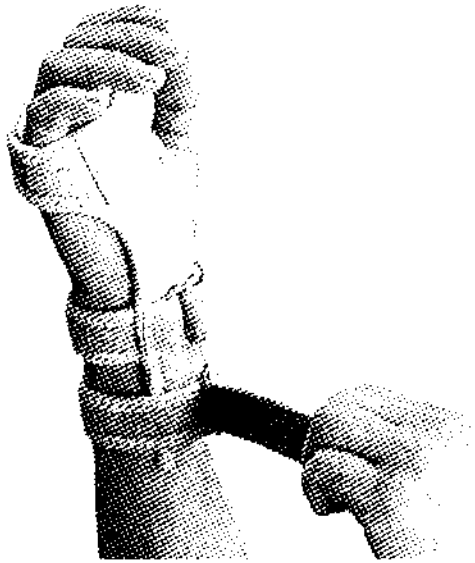


Fig. 45.6 Carpal tunnel splint

LOWER LIMB ORTHOTICS

A lower limb orthosis is an external device applied or attached to a lower body segment to improve function by controlling motion, providing support through stabilizing gait, reducing pain through transferring load to another area, correcting flexible deformities, and preventing progression of fixed deformities. A lower limb orthosis should be used only for specific management of a selected disorder. The orthotic joints should be aligned at the approximate anatomic joints. The orthosis selected should be simple, lightweight, strong, durable, and cosmetically acceptable.

Terminology

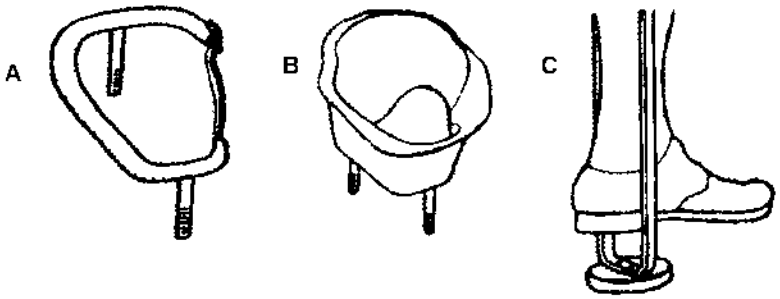
A logical and easy to use system of standard terminology has been developed, i.e., first letter is the name of the joint which the orthoses crosses and supports in proximal to distal sequence and the letter O for orthoses is attached at the end (Box). Orthoses generally are named by the body regions that they involve, as demonstrated by the following abbreviations:

AFO	Ankle-foot orthosis
KAFO	Knee-ankle-foot orthosis.
HKAFO	Hip-knee-ankle-foot orthosis.
THKAFO	Trunk-hip-knee-ankle-foot orthosis

Compound orthosis—Orthosis that cross more than five body segments, composed of two or more less complex devices (TLSO+HKAFO).
Hybrid orthosis—orthotic made of both metals and plastics.

Weight Bearing Devices

- **Ischial ring** (Figure 45.7) – Simpler to fabricate. Weight bearing surface is small. Little counterforce to maintain the ischial tuberosity on the ring.
- **Quadrilateral brim** (Figure 45.8) – gives good weight relief.
- **Patten bottom** (Figure 45.9) – for complete weight relieving with quadrilateral brim. Weight is transmitted directly by the orthoses from the ischial tuberosity to the patten bottom. Ankle joint is eliminated, lateral uprights extend distal to the shoe. Requires contralateral shoe lift.



A. Fig. 45.7 Ischial ring
B. Fig. 45.8 Quadrilateral brim
C. Fig. 45.9 Patten bottom

Shoes

- Forms foundation for orthosis
- Serves variety of functional and cosmetic purposes
- Minimizes pressure on sensitive, deformed structure
- Can be modified to redistribute weight towards pain-free areas to improve the comforts

Parts:
Bottom—insole, outsole, waist, heel.
Upper—vamp, quarters, toe cover

Ankle Foot Orthotics (AFO)

The position of the ankle indirectly affects the stability of the knee with ankle plantar flexion providing a knee extension force and ankle dorsiflexion providing a knee flexion force. An AFO is used in weakness or paralysis of ankle dorsiflexion, plantar flexion, inversion and eversion to prevent/correct deformities and reduce weight bearing.

Types

- Thermoplastic AFO
- Spiral AFO
- Hemispiral AFO
- Solid AFO: The solid AFO has a wider calf shell with trim line anterior to the malleoli. This AFO prevents ankle dorsiflexion and plantar flexion as well as varus and valgus deviation
- AFO with flange: This AFO has an extension (flange) that projects from the calf shell medially for maximum valgus control and laterally for maximum varus control.
- Hinged AFO: The adjustable ankle hinges can be set to the desired range of ankle dorsiflexion or plantar flexion.
- Metal and metal-plastic AFO: This type of AFO consists of a shoe or foot attachment ankle joint, two metal uprights (medial & lateral), with a calf band (application of force) connected proximally. The stirrup anchors the uprights to the shoes between the sole and the heel.

Special Purpose AFO

PTB Orthoses (Figure 45.10) - PTB brim replaces a calf cuff.

Fig. 45.10 PTB orthoses

Dennis Brown Splint (Figure 45.11)

Design: Consists of two plates that are securely attached to an infant's shoes and then connected to a cross bar. The footplates can be rotated on the cross bar to obtain a degree of rotation desired. Dorsiflexion of the ankles may be accomplished by appropriate bending of the cross bar.

Worn 24 hours a day in an attempt to correct rotation problems in the feet, tibia, or hips.

Used to hold the feet in varying degrees of dorsiflexion and hips in abduction.

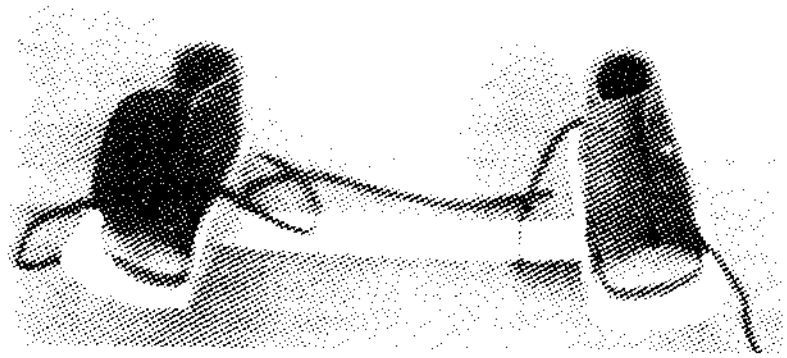


Fig. 45.11 Dennis Brown splint

Knee Ankle Foot Orthoses

is composed of an AFO with metal uprights, a mechanical knee joint, and two thigh bands. KAFO can be used in quadriceps paralysis or weakness maintain knee stability and control flexible genu valgum or varum. It its the weight bearing of the thigh, leg and foot with quadrilateral or ial containment brim.

Disadvantage

s more difficult to don and doff than an AFO, so it is not recommended patients who have moderate to severe cognitive dysfunction.

Modifications of KAFO

Scott-Craig Orthoses

sign

ible uprights, one posterior thigh band, a hinged pretibial band, ankle t with anterior and posterior adjustable pin stops, a cushion heel, a T ped footplate (embedded in the sole from heel to metatarsal heads).

Principle
Ankle joint in 10° of dorsiflexion is the key factor. The orthoses and the patients limbs thus lean forward slightly. Balance is achieved by compensatory hyperextension of the hip joints, so that the centre of gravity comes to a position posterior to the hip joints and anterior to the locked knee and ankle joints. Hinged pretibial band facilitates donning and doffing.

Application
Used in spinal cord injury patients for mobilization with orthotic stabilization of knee, ankle, and foot, passive or ligamentous stabilization to hip without additional components

Floor Reaction Orthoses

Principle
Applies forces that resists recurvatum and provide mediolateral stability. The distal portion limits subtalar motion and immobilizes the ankle in slight equinus so that a knee extension moment is produced when the forefoot is in contact with the ground. In the presence of knee extensor weakness, this orthoses eliminates the need for a mechanical knee lock during stance phase and permits knee flexion during swing.

Disadvantages
- Cannot be used bilaterally, since positioning both ankles in equinus interferes with anteroposterior stability.
- Proximal portion protrudes above the knee on sitting.

Modifications
Conventional knee joints and uprights are included to allow the prosthesis to flex at the knee joint while sitting.

Very durable because of its unitary construction and absence of mechanical parts.

Hip Knee Ankle Foot Orthosis (HKAFO) (Figure 45.12)

It is composed of a hip joint and pelvic band in addition to a KAFO.

Design: The orthotic hip joint is positioned with the patient sitting upright at 90°, while the orthotic knee joint is centered over the medial femoral condyle. Pelvic bands complicate dressing after toileting unless the orthosis is worn under all clothing. Pelvic bands increase the energy demands for ambulation.

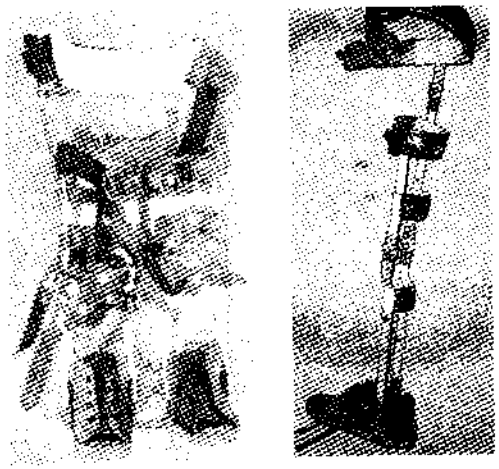


Fig. 45.12 Hip knee ankle foot orthosis (HKAFO)

Hip Joints and Locks (Figure 45.13)

- Single axis hip joints – allows flexion and extension but limited hyper extension. May include a lock to prevent flexion while standing, whether locked or unlocked it controls mediolateral (abd-add) motion.
- Double axis hip joints – allows lateral motion in addition to flexion and extension used where adduction and rotation control is desired.
- Double position hip locks – locking at both full extension and 90° of hip flexion. Useful for individuals with difficulty in maintaining the sitting position because of the spasticity of hip muscles.

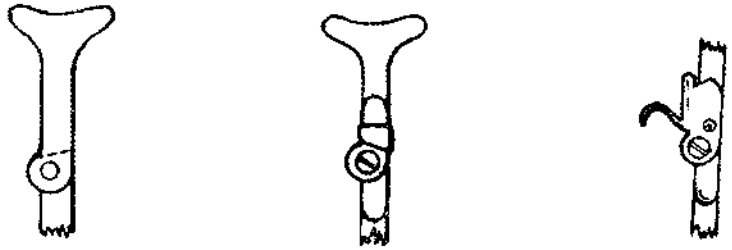


Fig. 45.13 Hip joints and locks

Trunk Hip Knee Ankle Foot Orthosis (THKAFO)

It consists of a spinal orthosis in addition to a HKAFO for control of trunk motion and spinal alignment. A THKAFO is indicated in paraplegics and very difficult to don and doff.

Pelvic Bands (Figure 45.14)

- Unilateral pelvic band – metal pelvic band encompassing the pelvis on the involved side below the iliac crest and above the greater trochanter, continues around the opposite side in the form of a flexible belt.
- Bilateral pelvic band – each side of the bilateral pelvic band begins at a point just posterior to the ASIS, midway between greater trochanter the iliac crests, then curves downwards and posteriorly to contact the most prominent portion of the buttocks and continues slightly upwards to overlie the sacrum.
- Double pelvic band – for maximum hip control.

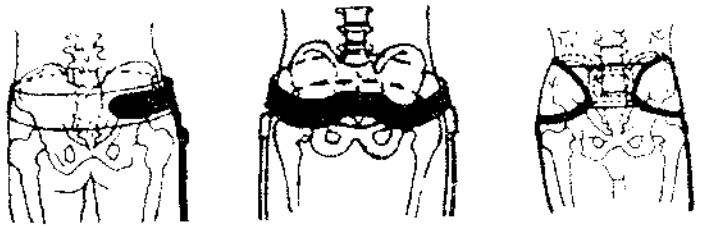


Fig. 45.14 Pelvic bands

Knee Orthosis (Figure 45.15)

It provides only support or control of the knee but not of the foot & ankle. The knee joint is centered over the medial femoral condyle. Because of short lever arm knee orthoses are not effective when strong forces are required and there is a tendency for these orthoses to slip.

- Knee orthoses for patello-femoral disorder: They are used to supply mediolateral knee stability and to control tracking of the patella during knee flexion and extension. E.g.: Infrapatellar strap KO, Palumbo KO.
- Knee orthoses for knee control in sagittal plane: They control genu recurvatum with minimal mediolateral stability. E.g.: Swedish knee cage, 3 way knee stabilizer.
- Knee orthoses for knee control in frontal plane: It consists of thigh and calf cuffs joined by sidebars with mechanical knee joints. The knee joint is polycentric and closely mimics the anatomic joint motion. E.g.: traditional metal-leather KO, Miami KO.
- Knee orthoses for axial rotation control: They provide angular control of flexion-extension and mediolateral planes, in addition to controlling axial rotation, being used in sports injuries of knee. E.g.: Lenox-Hill derotation orthoses, Lerman multiligamentous knee control orthoses.

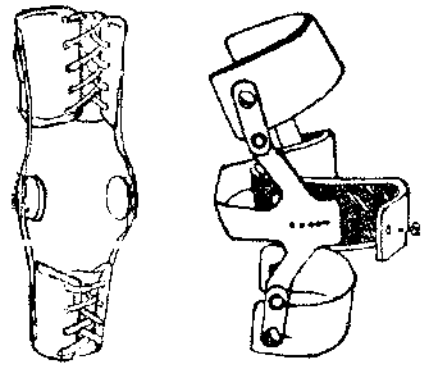


Fig. 45.15 Knee orthosis

SPINAL ORTHOTICS

An orthotic device (commonly just referred to as an orthotic) is an external device applied on the body to limit motion, correct deformity, reduce axial loading, or improve function in a certain segment of the body. Design characteristics of an orthotic device are crucial to function (Box).

Box 45.2 Important features of spinal orthoses

Functional use and adjustability
Cost and weight of material
Durability
Ability to fit various sizes of patients
Ease of putting on (donning) and taking off (doffing)
Access to tracheostomy site, peg tube, or other drains
Access to surgical sites for wound care aeration to avoid skin maceration from moisture
Cosmetic acceptance

Box 45.3 Indications for recommending orthotic devices

Pain relief
Mechanical unloading
Scoliosis management
Spinal immobilization after surgery
Spinal immobilization after traumatic injury
Compression fracture management
Kinesthetic reminder to avoid certain movements

Classification

- Supportive orthoses
  - Fabric supports—Belts & corsets
  - Rigid braces—Leather, plastic POP & plastazote.

2. Corrective orthoses
   - Acute corrective force in one or more directions

Box 45.4 Mechanism of pain relief by spinal orthoses

Psychological
Increases intra-abdominal pressure by abdominal compression
Supporting pendulous abdomen and thus decreases lumbar lordosis
Causes local inactivity of associated muscle groups and ligaments
Changes amount of movement occurring in different regions of spine

Duration of Orthotic Use

Duration of orthotic use is determined by the individual situation. In situations where spinal instability is not an issue, recommend use of an orthosis until the patient can tolerate discomfort without the brace. When used for stabilization after surgery or acute fractures, allow 6–12 weeks to permit ligaments and bones to heal.

Table 45.3 Relevant biomechanics of spine

Region	Properties
Cervical spine	Most mobile spinal segment of spine Flexion greater than extension Occiput and C1 also have limited side bending and rotation C1–C2 complex accounts for 50% of rotation in the cervical spine C5–C6 region has the greatest amount of flexion and extension C2–C4 region has the most side bending and rotation.
Dorsal spine	Least mobile Greater flexion than extension Lateral bending increases in a caudal direction, and axial rotation decreases in a caudal direction
Lumbar spine	Greatest movement in the lumbar spine is flexion and extension Minimal axial rotation

Box 45.5 The biomechanical principles in orthotic design

Balance of horizontal forces
Compression
Distraction
Construction of a cage around the patient
Placement of an irritant to serve as a kinesthetic reminder
Skeletal fixation

Cervical Orthotics

Soft Cervical Collar (*Figure 45.16*)

It is a made up of lightweight material, polyurethane foam rubber, with a stockinette cover. It has Velcro closure strap for easy donning and doffing. Anteriorly holes are provided for aeration. Comfortable to wear, but gets easily soiled with long-term use.

Indications

- Kinesthetic remainder to reduce motion and psychological comfort
- Support to the head during acute neck pain
- Relief with minor muscle spasm associated with spondylolysis

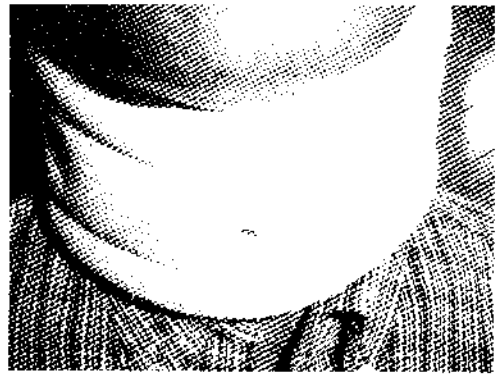


Fig. 45.16 Soft cervical collar

Hard Cervical Collar (*Figure 45.17*)

Devised by Hugh Owen Thomas. They are similar in shape to a soft collar but are made of plastizote, a rigid polyethylene material shaped like a ring with padding. Height can be adjusted in certain designs. The hard collar is more durable than a soft collar with long-term use.

Indications

- Stable fractures
- Post reduction of dislocations
- Whiplash injuries
- Torticollis
- Post operative rehabilitation.

Drawbacks of Cervical Orthotics

- The soft tissue structure compression around the neck (e.g., blood vessels, esophagus, trachea)
- Limits application of aggressive external force.
- The high level of mobility at all segments of the cervical spine makes it difficult to restrict motion.
- Cervical orthoses offer no control for the head or thorax; therefore motion restriction is minimal.
- Cervical orthoses serve as a kinesthetic reminder to limit neck movement.
- Continued long-term use has been associated with decreased muscle function.

Fig. 45.17 Hard cervical collar

Table 45.4 Salient features of cervial collars

Type of collar	Properties	Indications	Limitations of movements
Soft collar (Common orthotic device)	Lightweight material (olyurethane foam rubber, with a stockinette cover) Velcro closure strap for easy donning and doffing	Support to the head during acute neck pain Relief with minor muscle spasm associated with spondylolysis Relief in cervical strains	Flexion and extension by 5–15% Lateral bending by 5–10% Rotation by 10–17%

·ard cervical ·ollars	A rigid polyethylene material with padding Height can be adjusted Velcro straps for easy donning and doffing. More durable than a soft collar	Support to the head during acute neck pain Relief of minor muscle spasm associated with spondylosis Interim stability and protection during halo application	Better than a soft collar in motion restriction Full flexion and extension by 20–25% Less effective in restricting rotation and lateral bending

Philadelphia Collar *(Figure 45.18)*

·y are semi-rigid head-cervical orthotics with a two piece system of ·uzote foam. Plastic struts on the anterior and posterior sides are used ·upport. The upper portion of the orthosis supports the lower jaw and ·out, while the lower portion covers the upper thoracic region. An anterior · for tracheostomy is available. A thoracic extension can be added to ·se motion restriction and treat C6-T2 injuries. It is available in various · and is comfortable to wear with improved compliance and Velcro ·s for easy donning and doffing. However it is difficult to clean and ·mes soiled very easily.

·itions

- ·nterior cervical fusion
- ·lo removal
- ·ns Type 1 cervical fracture of C2
- ·nterior discectomy
- ·spected cervical trauma in unconscious patients
- ·ar drop fracture of the vertebral body
- ·rvical strain

Fig. 45.18 Philadelphia collar

Table 45.5 Salient features of head cervical orthotics

Type of orthoses	Properties	Indications	Movment restriction
Philadelphia collar	Semirigid with a 2-piece system of Plastizote foam Plastic struts on the anterior and posterior sides for support Upper portion supports the lower jaw and occiput Lower portion covers the upper thoracic region Velcro straps for easy donning and doffing. An anterior hole for a tracheostomy Difficult to clean	Anterior cervical fusion Halo removal Dens Type I cervical fracture of C2 Anterior diskectomy Suspected cervical trauma in unconscious patients Tear-drop fracture of the vertebral body Cervical strain	Flexion and extension by 65–70% Rotation by 60–65% Lateral bending by 30–35%
Miami J collar	Semi-rigid HCO Another cervical orthotic device 2-piece system made of polyethylene and a soft washable lining An anterior hole for a tracheostomy Velcro straps for easy donning and doffing	Same as above	Flexion and extension by 55–75% Rotation by 70% Lateral bending by 60%

Cervical Thoracic Orthotics

Sterno-Occipital-Mandibular-Immobilizer (SOMI) Brace *(Figure 45.19)*

This is rigid three-poster cervical thoracic orthoses with anterior chest plate that extends to the xiphoid process and has metal or plastic bars that curve over the shoulder. Straps from the metal bars go over the shoulder and cross to the opposite side of the anterior plate for fixation. A removable chin piece attaches to the chest plate with an optional headpiece that can be used when the chin piece is removed for eating. The SOMI is ideal for

bedridden patients since it has no posterior rods or back plate allowing the patient to lie flat on his back. It is more comfortable than a four-poster cervical brace. SOMI is less effective compared to other braces in controlling extension, but is very effective in controlling flexion at the atlantoaxial and C2–C3 segments.

Indications

- Immobilization in atlantoaxial instability because of rheumatoid arthritis. (ligamentous disruption affects flexion more & extension is held in check by the intact dens)
- Immobilization for neural arch fractures of C2 since flexion causes instability.

Fig. 45.19 Sterno-occipital-mandibular-immobilizer (SOMI) brace

Four-Poster Cervical Brace *(Figure 45.20)*

It is a modification of SOMI brace, consisting of padded chin and occipital supports attached by four adjustable turnbuckles to two plates anteriorly and posteriorly. Two shoulder and axillary straps connect the anterior and posterior plates.

Advantage: It does not interfere with radiological examination.

Fig. 45.20 Four-poster cervical brace

Table 45.6 Examples of cervical thoracic orthotics

Type	Salient features	Salient features	Movement restriction
Sternal-occipital-mandibular-immobilizer (SOMI)	Rigid three-poster CTO Anterior chest plate that extends to the xiphoid process Metal or plastic bars that curve over the shoulder Ideal for bedridden patients since it has no posterior rods	Immobilization in atlantoaxial instability	Cervical flexion and extension by 70%–75% Lateral bending by 35% Rotation by 60–65%
Four-poster brace	Rigid orthosis with anterior and posterior chest pads connected by a leather strap Molded occipital and mandibular support pieces connect to the chest pads and have adjustable struts	Immobilization in atlantoaxial instability	Flexion and extension by 80% Lateral bending by 55%–80% Rotation by 70%

Extended Cervical Collar (*Figure 45.21*)

It is made of rigid thermoplastic composed of two shells connected to each other by adjustable straps. Anteriorly it extends up to mandible superiorly and xiphisternum inferiorly. While posteriorly extends above the occiput.

Fig. 45.21 Extended cervical collar

Halo Device

The halo device is the most common device for treatment of unstable cervical and upper thoracic fractures and dislocations as low as T3.

Salient Features

- Provides maximum motion restriction of all cervical orthotics.
- Halo ring is made of graphite or metal with pin fixation on the frontal and parietal-occipital areas of the skull.
- Halo ring attaches to the vest anteriorly and posteriorly via 4 posters.
- The vest has shoulder and underarm straps for tightening made up of rigid polyethylene and extends down to the umbilicus.

Indications

- Odontoid Type I, II, and III fractures of C2
- Hangman fractures with rupture of the transverse ligament
- Atlantoaxial instability from rheumatoid arthritis
- Posterior neural arch fracture and disc disruption between C2 and C3
- Cervical tumour resection in an unstable spine
- Debridement and drainage of infection in an unstable spine

Contraindications
Concomitant skull fracture with cervical injury
Lacerated or infected skin over pin insertion sites
Cervical instability with ligamentous disruption
Cervical instability with 2 or 3 column injury

Motion Restrictions
Flexion and extension by 90–96%
Lateral bending by 92–96%
Rotation by 98–99%

Complications

Neck pain or stiffness Pin loosening Pin site infection Pin site pain pressure sores	Redislocation Neurological deterioration Avascular necrosis of the dens Ring migration

Important Considerations
Restriction in cervical motion depends on the fit of the halo vest.
Improper fit can allow 31% of normal spine motion.
Multidirectional shear forces can cause increased pinhole size with crater-like enlargement.
Device is used for 3 months to allow adequate time for bone healing.
Nearly 75% of patients without facet joint dislocation achieve good anatomic results.
Thorough neurologic examination before and after application of device.

Table 45.7 Efficiency of orthotic devices in controlling neck movements

Movement	Efficiency
Flexion and extension at C1–C3	Halo> four-poster brace> Cervicothoracic orthotics
Flexion and extension at C3–T1	Cervicothoracic orthotics
Flexion from C1–C5	SOMI brace
Rotation and lateral bending from C1–C3	Halo>cervicothoracic brace
Lateral bending	Four-poster brace>cervicothoracic brace

Thoracolumbar Orthotics

Taylor's Brace

In *1863 CF Taylor* described a rigid brace for Koch's spine treatment, consisting of metallic padded skeleton posteriorly, anteriorly shoulder straps superiorly and corset with abdominal straps inferiorly. It immobilizes T7 to L12 vertebrae primarily.

Advantages: Limits forward flexion, extension & lateral flexion of thoracolumbar region.

Disadvantages: Increases movement at lumbosacral junction.

Anterior Spinal Hyperextension (ASH) Brace

Prescribed by *Hoadley in 1896,* uses the principle of three point action of a bending force to extend the spine. It is made of padded thermoplastic anterior pieces superiorly over the sternum, inferiorly over the symphysis pubis and laterally over the mid axillary line with a padded posterior pad.

Indications
Wedge compression fractures of lower thoracic and upper lumbar spine.

Milwaukee Brace (Figure 45.22)

It is a CTLSO originally designed by *Walter Blount & Schmidt in 1958* for effective correction in ambulant treatment of structural scoliosis. It is also occasionally used for ankylosing spondylitis, tuberculosis or infections of upper thoracic spine.

Design: Consists of a leather corset ending superiorly just below the costal margin. Metal uprights, one anterior and two posterior pass to the neck to connect to the cervical ring. The ring is inclined 20° anteroinferiorly. Two occipital and one throat pad are provided for support. Rib rotations corrected with pressure pads over rib prominences. It is used for curves with apex above T7.

Indications
Patients with Risser score of I–II and curves greater than 20–30 degrees that progress by 5° over one year need brace application.

Curves between 30–40 degrees need bracing but not curves less than 20°.

Disadvantages
- Low deformity
- Pain
- Skin breakdown

- Unsightly appearance
- Difficulty with mobility and transfers
- Increase energy expenditure with ambulation

Boston Brace

Designed by *Hall & Miller in 1974* for lumbar and thoracolumbar scoliosis in which the apex of the curve is below the T8 vertebra. It is prefabricated symmetric thoracolumbar-pelvic mold with built-in lumbar flexion that can be worn under clothes.

Disadvantages

- Local discomfort
- Hip flexion contracture
- Trunk weakness
- Increased abdominal pressure
- Accentuation of hypokyphosis above brace in the thoracic spine

Knight's Brace

It consists of a metallic skeleton posteriorly with two vertical bars contoured to the lumbar lordosis and padded adequately. Anteriorly it has an abdominal corset superiorly, which extends to thoracolumbar junction and inferiorly upto sacrococcygeal junction.
It is indicated for immobilization of midlumbar spine.

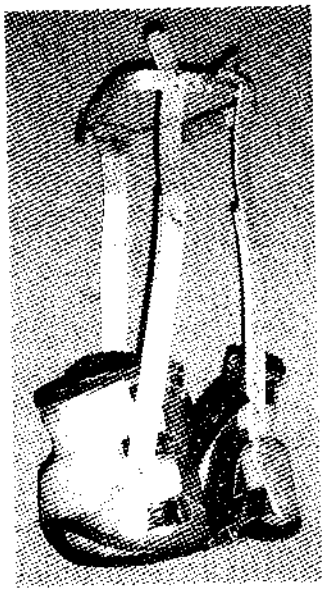


Fig. 45.22 Milwaukee brace

Lumbosacral Corset (*Figure 45.23*)

These are fabric supportive orthosis. It provides subjective support and kinesthetic remainder. It extends upto thoraco-lumbar junction posteriorly and covers abdomen anteriorly. Posteriorly there is a rigid metal strip to reinforce the support. It probably acts by increasing intraabdominal pressure and thus decreases pressure on the spine.

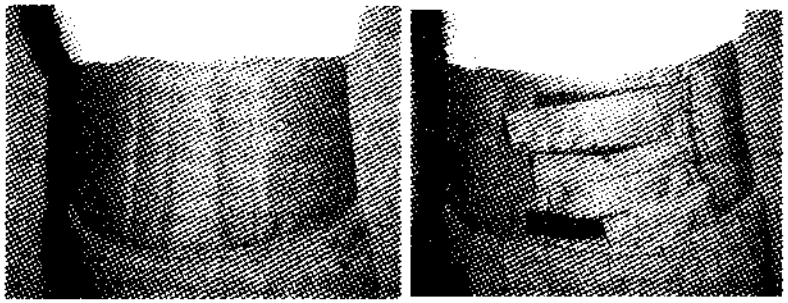


Fig. 45.23 Lumbo-sacral corset

APPLIANCES FOR PARALYSIS

Calipers

The calipers are used mainly for the flail lower limbs (e.g., polio), and to give support to otherwise weak and unstable knees and ankles.

Types of Calipers

Below-knee Caliper

The main components of below-knee caliper are galvanised wire or soft iron or mild steel. Those made out of the latter metal is more difficult to bend but is stronger. The top of the ring is padded with wool or cotton waste, and covered with leather as shown. Soft lining leather is used next to the skin.

Above-knee Caliper

This is identical with a below-knee caliper except for its length, the size of the ring, and has a simple knee-piece to support the knee. In the case of genu recurvatum, a tight posterior strap a little wider than normal and prevents the knee from displacing backwards.

Hip Flexion Pieces

In patients with very unstable hips, especially those with an internal or external rotation deformity, a simple hinge on a pelvic band will improve it. In those with a very unstable spine this is attached to the corset, instead of to a simple pelvic band.

Knee Bending Caliper
This has the advantage of being adjustable for a growing child.

Clog with a Backstop

A foot with a tendency to a foot drop is most easily corrected by a simple clog with a backstop. This backstop, if reversed, can also be used to prevent a calcaneus foot occurring.

Crutches and Walking Aids

Crutches are essential for patients with above-knee calipers on both knees. These patients often have weak hips and lower trunk muscles as well. The only method of propulsion forward is by the use of crutches.

Except for special cases the patient must fulfill the following criteria before crutches are prescribed:
1) A grip strong enough to hold the crutch
2) Triceps strong enough to propel himself forward
3) A spine strong enough to allow the patient to sit up unaided unless the arms are very powerful.

Simple Wooden Crutch

It has the disadvantage, however, of having a fairly narrow base for patients who are very unstable, and it is not as strong as a well made metal crutch. It can be made adjustable by providing several holes for the hand-hold and a through-bolt and a wing nut. Length can be adjusted by making a sliding base which can be elongated and held by two bolts with wing nuts.

Metal Crutches

This has the advantage over the wooden crutch of durability and stability. It has the disadvantages of being heavier, more expensive and much more difficult to make.

Walking Sticks

Many patients with a single paralysed leg can be managed with a single stick. It is essential that the stick is held in the hand of the side opposite to that of the weak or weaker leg, and not on the same side.

Simple Parallel Bars

Parallel bars will allow a patient who cannot quite manage to walk with crutches to learn how to walk. It will also enable those who can walk slowly to walk better.

CHAPTER 46

Wheel Chair

Before prescribing the wheel chair, therapists must have specialized knowledge of the various types of disabilities; the short-term and long-term prognoses; neuromuscular assessment of seated functioning, including range, tone, and reflexes; biomechanical principles of support; therapeutic options and outcomes; and other medical interventions that may occur in the future.

SECTION OF WHEEL CHAIR

Wheel chairs which are cheap, strong and can be locally manufactured, are required for patients with severe poliomyelitis, patients with paraplegia and quadriplegia due to spinal tuberculosis, injuries and transverse myelitis. There are also numerous patients with severely deformed or amputated limbs due to trauma, untreated congenital deformities, severe osteomyelitis, pyogenic and tuberculous arthritis and leprosy.

GOALS OF A WHEELCHAIR SYSTEM (BOX 46.1)

Goals of a wheelchair system include facilitated mobility, positioning, support, adaptations to temporary or permanent conditions, and optimization of function. An appropriately used system that is accepted by the patient and caregiver is an indicator of a well-developed system.

Box 46.1 Goals to achieve while prescribing wheel chair

- Mobility and safety
- Decrease tone influences
- Support to prevent deformity
- Prevention of pressure sores
- Provision of stability to enhance functional control of the head or extremities
- Support for the trunk to compensate for muscle weakness
- Increase in sitting tolerance
- Relief of stress on painful or injured areas of the body
- Accommodate deformities and joint limitations

PATIENT INFORMATION

Age and sex
Cognitive status
Visual/auditory status
Past medical/surgical history
Definie the present problem (i.e., detailed physical and neurological examination and postural assessment)
Anticipation of future problems (i.e., bed sores)
Underlying condition static or progressive
Likely course of the disability
Patient's functional performance assessement

WHEELCHAIR CONSIDERATIONS

Does the patient require a cushion that increases sitting balance to free the arms for functional activities?
How should the patient be positioned for optimum access to the wheels of the chair?
How does the patient transfer?
What seat height is the safest and most functional?
Will the weight of the cushion affect the patient's ability to propel the chair?
Is the patient in a position to adjust, maintain, or replace the cushion on a regular basis?

Functions Assessment

Home access
Type of transfer skills; assist level
Propulsion – manual versus powered
Activities of daily living (ADL) skills

Tray, communication device, and environmental control
Bowel and bladder functions

Posture Assessment

Pelvis – Tilt/obliquity
Hip adduction/abduction tendencies
Back – Alignment; fixed or flexible deformities
Neck – Posture, alignment, and control
Ankle – Position
Skin – Areas of increased risk (e.g., spine, coccyx, ischial tuberosities, sacrum)
Tone – Reflexes (flexor/extensor and symmetric and asymmetric tonic neck)
Body habitus – Height, weight, and girth
Any equipment – Baclofen pump, shunts, gastrostomy tubes, lines, ventilator

CHAPTER 47

Preoperative Assessment and Management

Preoperative evaluation in any patient should include history, physical examination, laboratory tests, and an assessment of surgical risk to identify coexisting diseases and complicating conditions and to correct the few that are reversible. Before emergency surgery, complete evaluation and correction of physiologic abnormalities may be impossible, but obtaining as much information as possible helps the surgeon take precautions to prevent complications.

INFORMED CONSENT

Before surgery, the surgeon explains the procedure, including possible complications, to the patient. If a patient has dementia or delirium and cannot understand the surgical risks and benefits, the surgeon must proceed with care based on decisions made by a surrogate.

NURSING ISSUES

Nurses should teach patients about what to expect during recovery. As soon as possible, often hours after surgery, patients should get out of bed and be mobile; an active rehabilitation program is developed to help patients quickly regain function.

HISTORY

The surgeon should obtain the patient's history before surgery. The accuracy of the history in neurosurgical patients may be limited by sensory or cognitive deficits, requiring the surgeon to rely on a history given by caregivers.

PREOPERATIVE EVALUATION

Age

The percentage of patients with coexisting diseases increases dramatically with age, from < 30% among patients aged 21 to 30 years to 90% among those aged 71 to 80 years.

General Health Status

The American Society of Anesthesiologists Physical Status Classification is commonly used to predict surgical outcome based on a patient's preoperative health status. The mortality rate for different age groups varies little among patients in classes I and II and only slightly among those in classes III and IV when grouped by age.

The American Society of Anesthesiologists Physical Status Classification

- Class 1 : Healthy
- Class 2 : Mild systemic disease
- Class 3 : Severe but not incapacitating disease
- Class 4 : Severe disease, constant threat to life
- Class 5 : Moribund patient not expected to survive for 24 hours

Functional Status

All complications, including life-threatening ones, are more common in inactive patients than in those with normal activity levels.

Nutritional Status

The mortality rate is significantly higher in patients with malnutrition.

Psychologic Status

Dementia is a major risk factor for a poor outcome and also postoperative complications are more common in demented patients.

Box 47.1 Systemic illnesses that increases surgical risk

Cardiovascular diseases
Myocardial infarction and heart failure
Cardiac arrhythmias
Hypertension
Carotid artery occlusive disease
Pulmonary disease
Asthama
Bronchitis
Impaired liver functions
Coagulation abnormalities
Renal disease
Acute or chronic renal failure

Physical Examination

A complete physical examination should include examination of the skin, oral mucosa, and tongue. It will provide information about hydration and nutrition. Femoral, popliteal, and pedal pulses should be noted, and evidence of venous disease (e.g., varicose veins, postphlebitic ulcers, oedema) identified. Detail neurological examination should be noted.

Laboratory Tests

Urinalysis, a peripheral blood count, chemistry tests, and measurement of coagulation factors are necessary. A chest X-ray and an ECG are routinely obtained.

INTRAOPERATIVE MONITORING

Intraoperative monitoring of blood pressure, heart rate and rhythm, ventilation, fluid status, and urinary output is performed in all patients. Other parameters needs to be monitored are oxygen saturation and body temperature.

POSTOPERATIVE CARE

All patients are treated in an intensive care unit or recovery room for < 72 hours before being returned to a standard hospital room.

Complications

Early Postoperative Complications

Hypoxaemia

Analgesics, particularly use of opioids, can cause hypoxaemia in post-operative period. It can be treated with supplemental oxygen until arterial saturation is similar to the preoperative level.

Pain

Controlling pain is as important as maintaining appropriate blood pressure and body temperature in post-operative period.

Postoperative Delirium

Postoperative delirium is characterized by difficulty in organizing and coordinating thoughts and by slowed motor function. It ranges from mild confusion to full hallucinations. Use of certain anaesthetics, meperidine, and anticholinegics increases the risk of postoperative delirium. Other causes of delirium are fluid and electrolyte imbalance, drugs, sleep deprivation, frequent interruptions for nursing care, altered circadian rhythms, and an inability to keep track of time.

Hypotension

The most common cause of hypotension during the early postoperative period is hypovolaemia due to inadequate replacement of intraoperative fluid losses, occult haemorrhage, or internal fluid losses (e.g., reaccumulation of ascites, third-space losses).

Hypothermia

Immediately after a long operation, body temperature may decrease to 32.2° to 35.0°C (90° to 95°F). Treatment includes warming IV and other fluids and using convection warming systems.

Respiratory Problems

These include aspiration, atelectasis and pneumonia.

Fluid and Electrolyte Imbalance

During the early postoperative period, the body normally retains water and sodium, and the person may have difficulty eliminating the excess fluid. This can lead to combination of electrlyte disturbances (e.g., hypernatremia, hypokalaemia etc.).

Nutritional Deficiencies (Discussed in Chapter 44)

Late Postoperative Complications

Pressures sores
Fat embolism
Pneumonia
Cardiac complications (e.g., heart failure, arrhythmias)
Septicaemia

CHAPTER 48

Surgical Considerations and Reconstruction

CONTRACTURES (FIGURE 48.1)

Contractures will usually occur if there is imbalance of single group of muscles which are not held in check. Other factors responsible for contracture formation include the effects of gravity, the effects of flexed joints in bed, and the results of bearing weight on a leg. Secondary to the primary causes are the effects on joint, bone connective tissue of the initial contracture. The joint capsules become contracted on the flexed side, the epiphysis may become flattened or deformed, and intermuscular septa, nerves and vessels become shortened in time. The skin over the flexed face may also become tight. Knee and ankle contractures may prevent the fitting of calipers.

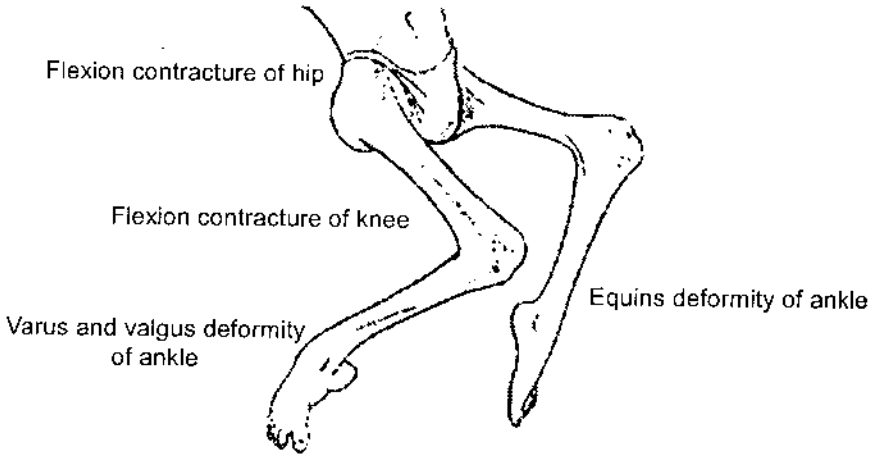


Fig. 48.1 Contractures in the lower limbs

Prevention of Contractures

The prevention of contractures in the acute and subacute stages is very important. Treating therapist should know how to stretch the paralysed limbs daily to prevent contractures.

Splints

Splints, except for back slabs for a drop foot, are now seldom used. Reliance is placed much more on daily stretching plus correct support of paralyzed limbs in bed.

Stretching of Muscles and Joints (Figure 48.2, 48.3 and 48.4)

Joints must be stretched in the direction opposite to that of the contracture, i.e., an equinus ankle dorsally. This must be carried out at least once a day by the physiotherapist or nurse, and at least three times a day by relatives. The important contractures are those of hip, knee and ankle, but other contractures such as a varus contracture of the foot, or an adduction contracture of the shoulder may occur.

Flexion Contracture of the Hip (Figure 48.2)

In manipulation of the hip, the pressure backwards should be in the upper third of the thigh, lest excessive leverage should cause a fracture. The opposite hip must be fully flexed to eliminate lumbar lordosis, and the leg should be brought down in slight adduction to stretch the abductors which are usually also tight. Lying the patient on his face in bed, with a pillow under the lower thigh is useful, provided the patient will tolerate the position. The hips can also be extended while the patient is in this position.

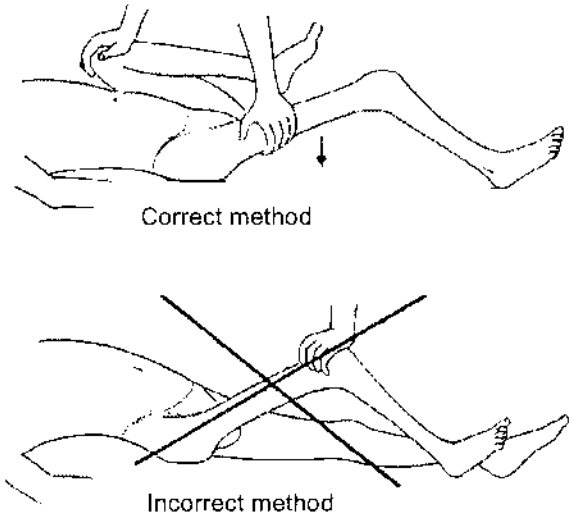


Fig. 48.2 Correct and incorrect method of hip manipulation

Flexion Contracture of Knee (Figure 48.3)

It is essential that the knee is manipulated as shown in the figure, and that pressure is exerted near the joint. If this is not done, fractures of the tibia or femur plus slipping of the epiphyses and backward subluxation of the tibia on the femur are liable to occur.

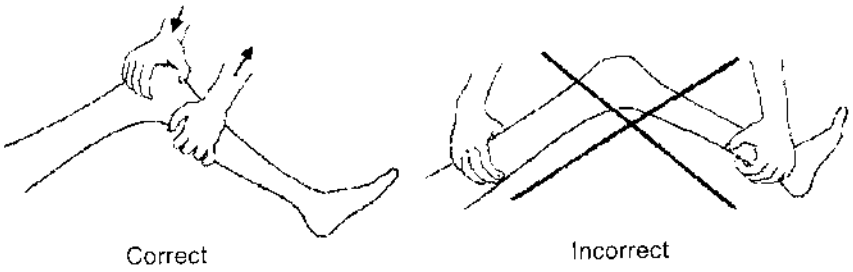


Fig. 48.3 Correct and incorrect method of knee manipulation

Preventing a Recurrence of Contracture

Apart from stretching imbalanced muscles, the only way of preventing a recurrence of a contracture is to hold a joint in an overcorrected position. This is so the deforming muscles are acting at a mechanical disadvantage. This is most easily achieved by fitting calipers as soon as the tender muscles will allow, and leaving the calipers on for most of the day and night in the acute and subacute stages.

Manipulation of Ankle and Foot Deformities (Figure 48.4)

This is shown, and the most important deformity to correct is equinus. The correct method is demonstrated with the ankle firmly supported as the foot is dorsiflexed. In the case of varus of the foot, or adduction of the forefoot, it is important to be firm yet gentle, and to avoid too rapid or forceful a manipulation. Much more is achieved by firm pressure for at least five minutes in the opposite direction to the deformity. This will usually need to be repeated, and followed by surgical correction to prevent recurrence.

Hip and Knee Ccontractures

It is essential that the patient is assessed in detail before any operative procedures are considered. The main criteria with any operation on the lower limb are whether the patient is likely to be able to walk with or without a caliper following an operative procedure, and whether the patient will be socially benefited by being able to walk. Hip and knee contractures of over 30° will require operative correction.

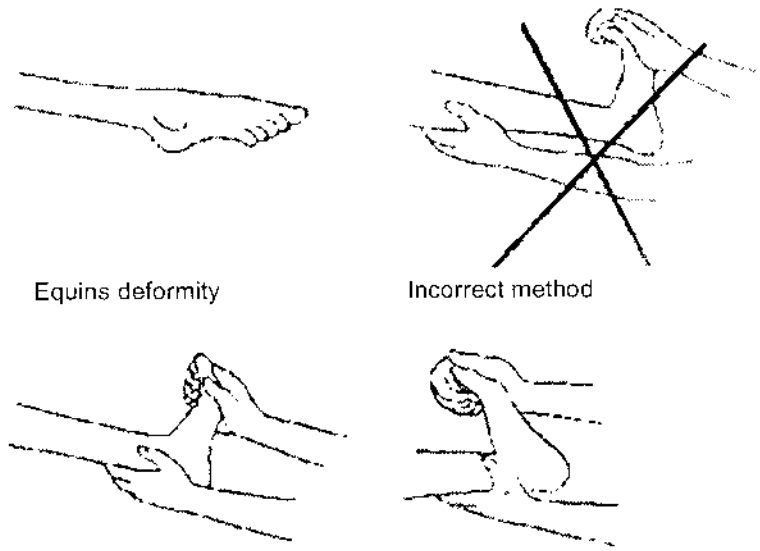


Fig. 48.4 Correct and incorrect methods of ankle manipulation

Isolated Hip Contractures of Less than 30°

No treatment is required for these when there are no other contractures. The stability of the hip is often improved, and shortening compensated for, if there is a small degree of abduction/flexion contracture.

Isolated Knee Contractures of Less than 30°

In Children

In a child these are best treated by fortnightly manipulations under anaesthesia until at least 2° of genu recurvatum is obtained. An above knee caliper is then fitted. If there is associated hip contracture preliminary soft tissue correction will also be necessary. Russell traction will also correct this deformity but will take time and will necessitate hospitalisation.

In Adults

In adults this contracture may be much more difficult to correct. Manipulation alone usually fails. Russell traction again takes time but may be effective. In some adult patients with a mild degree of flexion deformity no treatment however is necessary. In others a supracondylar osteotomy may be indicated, but only if the foot is stable and a caliper may thereby be discarded.

UPPER EXTREMITY

The results of surgery on the upper extremity, in which the goal is functional mobility, are poor compared with those of the lower extremity, in which the goal is painless stability. Operations on the upper extremities are designed primarily to place the arm and forearm in a functional position and to enable the patient to extend the fingers and wrist while retaining active flexion of the fingers.

Shoulder

Contracture of the shoulder or spasticity of the muscles that control it usually is not disabling enough to justify surgery.

Adduction and Internal Rotation

Neurectomy of motor nerves to the involved muscles is impractical because the nerves are not easily accessible.

- Fairbank operation
- Rotational osteotomy of the humerus at the level of the deltoid tubercle

Elbow

Elbow Flexion Contracture

Release of elbow flexion contracture

LOWER EXTREMITY

Deformities of the Foot

Equines
Varus
Valgus
Talipes calcaneus
Adduction of the forefoot
Hallux valgus with bunion formation
Clawing of the toes

Talipes Equinus

Conservative treatment often is sufficient, but sometimes the deformity is so severe that surgery is indicated. In a young child the deformity often can be corrected conservatively by stretching the triceps surae both manually and by using a brace. Inhibitive casts that are supposed to obliterate the plantar reflex are actually excellent holding casts that allow the stretched gastrocsoleus complex to lengthen.

Surgical Correction

Surgery is indicated in equinus deformity when conservative treatment fails or when the deformity is so severe that conservative treatment would be ineffective. Because the equinus deformity tends to recur until growth is complete, the patient must be monitored until skeletal maturity.

Tendon lengthening procedures

- Open lengthening of tendo calcaneus
- Percutaneous lengthening of tendo calcaneus
- Semiopen sliding tenotomy of tendo calcaneus
- Lengthening of gastrocnemius muscle

Muscle Transfer Techniques

Aim of these techniques is to achieve dorsiflexion

- Flexor hallucis longus and flexor digitorum longus transfer to the dorsum of the foot after tendo calcaneus lengthening
- Anterior transfer of long toe flexors for spastic equinus and equinovarus

Tendon transfers require prolonged physiotherapy post-operatively, and are seldom indicated as isolated procedures.
Neurectomy of branches of tibial nerve to the gastrocnemius or soleus, or both, with advancement of the insertion of the tendo calcaneus.
Anterior advancement of tendo calcaneus

Talipes Equinovarus

The goal of surgery should be to render the patient free of bracing or to improve walking in a brace when proprioception is defective or the dorsiflexor muscles are inadequate.

Indications for Surgery

To improve function when standing and walking
To aid in fitting the patient with shoes
To correct deformity when an orthosis will not hold it corrected
To allow the extremity to be used without the impairment of an orthosis

Surgical Procedures

Lengthening of tibialis posterior tendon
Z-plasty procedure
Step-cut procedure
Sliding lengthening at its musculotendinous junction
Intratendinous lengthening of the tibialis posterior tendon

Talipes Calcaneus

Usually is secondary to excessive or repeated lengthening of the tendo calcaneus either alone or in conjunction with neurectomy of branches of the tibial nerve.

It also can develop as a primary deformity when the dorsiflexors of the foot are spastic and the triceps surae is weak.

Surgical Correction

Partial denervation of toe extensors and transfer of the tibialis anterior tendon tendo calcaneus

Insertion of peroneus longus and tibialis posterior tendons into the tendo calcaneus to strengthen plantar flexion.

Crescentic osteotomy of calcaneus

Crescentic osteotomy of the calcaneus is to lengthen the foot and elevate the base of the heel.

Talipes Cavus

Hindfoot Cavus

Crescentic osteotomy of the calcaneus combined with plantar release

Forefoot Cavus

Plantar release and casting

Adduction deformity of forefoot

Caused by spasticity of the abductor hallucis muscle

- Passive correction
- Resection of a segment of the muscle and its tendon

Hallux Valgus

Usually occurs secondary to equinovalgus in the foot, valgus in the heel, or external torsion of the tibia.

Correction of the equinovalgus, heel valgus, or external tibial torsion should precede any bunion procedure.

***Claw Toes** (Figure 48.5)*

Neurectomy of the lateral plantar nerve

Metatarsophalangeal joint capsulotomies and tenotomy of the long toe extensors to the lesser toes and proximal interphalangeal joint resections or fusions using Kirschner wire fixation until the bone and soft tissues are stable.

Knee

***Knee Flexion Deformity** (Figure 48.6)*

Most common knee deformity

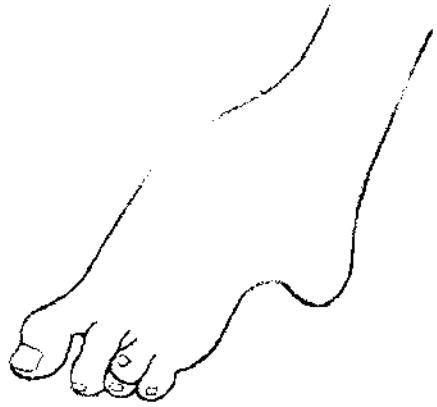


Fig. 48.5 Claw toes

Causes

Tight hamstring muscles, weak quadriceps muscle, or a combination of both.

Hip flexion deformities in which the hip flexors are spastic, the hip extensors are weak, or a combination of the two.

Equinus of the ankles, in which the hips are flexed, the knees are flexed, and the ankles are in equines.

Secondary to a weak triceps, in which the tendo calcaneus or another portion of the triceps surae has been overlengthened.

Surgical Procedures

- Fractional lengthening of hamstring tendons
- Distal transfer of rectus femoris
- Lateral transfer of medial hamstrings for internal rotational deformity of hip

***Recurvatum of Knee** (Figure 48.7)*

Genu Recurvatum

Causes

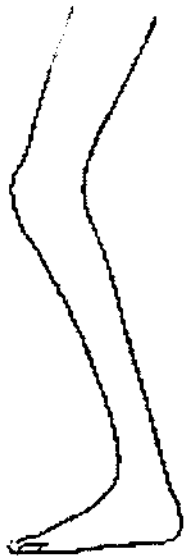
Quadriceps spasticity

Hamstring weakness

Gastrocnemius muscle weakness

Treatment

In children this should be treated by an above knee caliper with a tight posterior strap if it is more than 10°. In adults it should be left untreated if it is not getting worse or causing complications. A corrective osteotomy is occasionally required.



Knee flexion deformity

Fig. 48.6 Knee flexion deformity

Genu recurvatum

Fig. 48.7 Genu recurvatum

Knee Valgus (Figure 48.8)

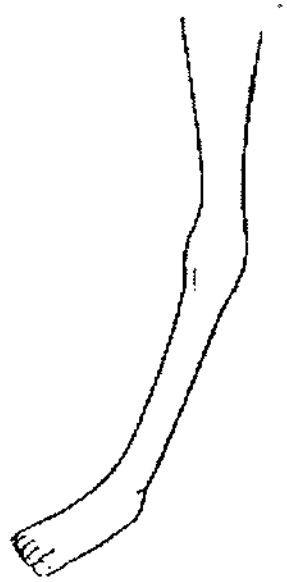
This is common and seldom requires specific treatment except for adjustment of a caliper to prevent it rubbing. In severe cases an additional valgus knee strap can be used.

Causes

Hip adduction deformity, coupled with internal rotation and flexion

Surgical Procedure

Correction of the spastic hip adduction and internal rotational deformity.
If the hip adductors, the medial hamstrings, or the iliopsoas cause the deformity than cause must be determined and corrected surgically.
Supracondylar varus osteotomy for severe deformity
Subluxation or dislocation of patella
Results from a valgus knee deformity
Can be caused by flexion, adduction, and internal rotation contracture of the hip.



Genu valgum

Fig. 48.8 Genu valgum

Surgical Procedure

- Reduction of dislocated patella
- Transferring the patellar tendon insertion
- If there is a valgus knee deformity than a varus supracondylar osteotomy of the femur may be needed.

Hip

Dislocation or Subluxation of the Hips

This is occasionally seen with severe paralysis of the hip muscles. It is more usual, however, after an extensive open division of the hip muscles and ligaments for a flexion deformity. Dislocations require reduction and treatment by an abduction spica. Some cases of recurrent dislocation are best left untreated. Others may require an osteotomy, an arthrodesis, or occasionally a psoas transfer.

Causes

Imbalance of muscle power
Retained primitive reflexes
Habitually faulty posture
Absence of weight-bearing stimulation on bone, and growth

Adduction Deformity

Scissoring gait and subluxation and dislocation of the hip

Surgical Procedures

- **Adductor longus tenotomy** to prevent further subluxation and dislocation of the hip
- **Iliopsoas recession**
- Adductor tenotomy and anterior obturator neurectomy
- Transfer of adductor origins to ischium

Tendon Transfers

Tendon transfer shifts a tendinous insertion from its normal attachment to another location so that its muscle can be substituted for a paralyzed muscle in the same region. Tendon transfers are indicated when dynamic muscle imbalance results in a deformity that interferes with ambulation or function. Surgery should be delayed until the maximal return of expected muscle strength in the involved muscle has been achieved.

Objectives of a tendon transfer

1) To provide active motor power to replace function of a paralyzed muscle or muscles.
2) To eliminate the deforming effect of a muscle when its antagonist is paralyzed.
3) To improve stability by improving muscle balance.

The ideal muscle for tendon transfer would have the same phasic activity as the paralyzed muscle, would be of about the same size in cross section and of equal strength, and could be placed in proper relationship to the axis of the joint to allow maximal mechanical effectiveness. Unfortunately, not all of these criteria can be met in every instance.

Section VII

MISCELLANEOUS

CHAPTER 49

Considerations in Neuro-rehabilitation

CONCEPT OF REHABILITATION

The central concept in rehabilitation is to foster the natural recovery from illness, to avoid adding iatrogenic problems, and to prevent and treat secondary complications of disease and its treatment.

NEUROLOGICAL DISABILITY

Neurological diseases are a major cause of long-term disability in society. Head injuries, strokes, multiple sclerosis, cerebral palsy, and epilepsy are particularly common examples. Rehabilitation is beginning to explore what might be achieved in those with cognitive dysfunction resulting from brain damage. The choice between residential and home care depends upon the nature of the disability, the patient's social circumstances, and the local availability of facilities. Ideally, this care, and treatments aimed at minimizing disability, should be supervised by an advisory team led by a physician in neurological disability, which also includes speech and occupational therapists, a physiotherapist, a nurse, and a social worker. Each patient requires an individual rehabilitation programme, and each member of the team must know what is expected. It is vital to involve carers at every stage.

Box 49.1 Types/classification of disabilities

- Locomotor disabilities
- Visual disabilities
- Communication disabilities
- Mental retardation
- Cerebral palsy
- Mental illness
- Multiple disabilities (more than one disability in the same person)

CARE PLAN

For each patient a care plan is defined. This is based on a full diagnosis and knowledge of the likely pattern of recovery from illness, the patient's present level of functioning, and the demands that his or her home environment are likely to make. The care plan defines objectives, what treatment is to be offered to the patient, and what progress is to be expected. At regular intervals the patient's functional state is reviewed by the multidisciplinary team and compared with the original plan. If there is a mismatch, the reason should be sought. Has some new illness intervened? Has a complication of illness (such as reactive depression) arisen? Was the original assessment of probable progress unrealistic? On the basis of this review the plan is updated and revised if necessary. The overall strategy necessitates a realistic view of the patient and his or her psychological state, diseases and disabilities, an organized setting, and a co-ordinated method of management, usually involving a large number of points of detail. Although there may be little scientific evidence that rehabilitation as at present conducted affects eventual outcome (for example in stroke), it is highly unlikely that all its benefits are psychological, while the consequences of a lack of rehabilitation (for example contractures and unnecessary immobility) are obvious.

STAGES IN RECOVERING FROM ILLNESS

The process of recovering from any illness usually includes treatment, spontaneous recovery, rehabilitation, and the return to community living.

Treatment

Treatment for stroke begins in a hospital with "acute care." This includes helping the patient survive, preventing another episode, and taking care of any other medical problems.

Spontaneous Recovery

Spontaneous recovery happens naturally to most people. This process is quickest during the first few weeks, but it sometimes continues for a long time.

Rehabilitation

Rehabilitation helps the person keep abilities and gain back lost abilities to become more independent. Rehabilitation usually begins while the patient is still in acute care.

Community Living

This may include doing common tasks in new ways or making up for damage or limits of one part of the body by greater activity of another.

REHABILITATION PROGRAMS

The setting for rehabilitation varies according to the needs of the patient and, particularly, the level of rehabilitative therapy needed. The choice between residential and home care depends upon the nature of the disability, the patient's social circumstances, and the local availability of facilities. Ideally, this care, and treatments aimed at minimizing disability, should be supervised by a rehabilitation team. There are several kinds of rehabilitation programs:

Hospital Programs

Institutionalization may be necessary for patients with severe disabilities and also hospital programs are usually more intense than other programs and require more effort from the patient.

Nursing Home Programs

As in hospital programs, the person stays at the facility during rehabilitation. Some provide a complete range of rehabilitation services; others provide only limited services.

Outpatient Programs

Outpatient programs allow a patient who lives at home to get a full range of services by visiting a hospital outpatient department, outpatient rehabilitation facility, or day hospital program.

Home-based Programs

The patient can live at home and receive rehabilitation services from visiting professionals. Providing rehabilitation at home by family members is highly desirable, especially for patients with substantial financial resources but it can be physically and emotionally taxing for caregivers. Other important advantage of home programs is that patients learn skills in the same place where they will use them.

INDIVIDUAL REHABILITATION SERVICES

Many patients do not need a complete range of rehabilitation services. Instead, they may need an individual type of service, such as regular physical therapy or speech therapy. These services are available from outpatient and home care programs.

Treatment Planning

Each patient requires an individual rehabilitation programme, and each member of the team must know what is expected. It is vital to involve carers at every stage.

Box 49.2 Rehabilitation team

- Physicians
- Nurses
- Physical therapists
- Occupational therapists
- Speech therapists
- Psychologists
- Social workers
- The patient
- Family members

Because every patient is different, treatment will be different for each person. Rehabilitation is provided by several types of specially trained professionals. A person may work with any or all of these:

Physician

Several kinds of doctors with rehabilitation experience may have this role. These include family physicians and internists (primary care doctors), geriatricians (specialists in working with older patients), neurologists (specialists in the brain and nervous system), and physiatrists (specialists in physical medicine and rehabilitation). An urologist may help with bladder problems. Other physician specialists may help with medical or emotional problems.

Rehabilitation Nurse

Rehabilitation nurses specialize in nursing care for people with disabilities. They provide direct care, educate patients and families, and help the doctor to coordinate care.

Physical Therapist

Physical therapists evaluate and treat problems with moving, balance, and coordination. They provide training and exercises to improve walking, getting in and out of a bed or chair, and moving around without losing balance. They teach family members how to help with exercises for the patient and how to help the patient move or walk, if needed.

Occupational Therapist

Occupational therapists provide exercises and practice to help patients do things they could do before the illness such as eating, bathing, dressing, writing, or cooking.

Speech Therapist

Speech therapists help patients get back language skills and learn other ways to communicate. Speech therapists also work with patients who have swallowing problems (dysphagia).

Social Worker

Social workers help patients and families make decisions about rehabilitation and plan the return to the home or a new living place. They may also provide or arrange for patient and family counseling to help cope with any emotional problems.

Psychologist

Psychologists are concerned with the mental and emotional health of patients. They may also treat thinking or memory problems or may provide advice to other professionals about patients with these problems.

Therapeutic Recreation Specialist

These therapists help patients return to activities that they enjoyed before the illness such as playing cards, gardening, or community activities. Recreational therapy helps the rehabilitation process and encourages the patient to practice skills.

Orthotist

An orthotist may make special braces to support weak ankles and feet.

Dietitians

Dietitians make sure that the patient has a healthy diet during rehabilitation. They also educate the family about proper diet after the patient leaves the program. Vocational counselors may help patients go back to work or school.

Assessment of Disabilities

Each patient is different and extent of disabilities depends on the part of the central nervous system injured and extent of injuries. Some of the examples are listed below:

- Weakness (hemiparesis) or paralysis (hemiplegia, paraplegia or quadriplegia)
- Problems with balance or coordination
- Locomotor disabilities
- Communication disabilities—aphasia and dysarthria
- Bodily neglect or inattention—being unaware of or ignoring things on one side of the body
- Pain, numbness, or odd sensations
- Cognitive problems (Problems with memory, thinking, attention, or learning
- Dysphagia (difficulty in swallowing)
- Bowel or bladder dysfunction
- Mental illness—depression

REHABILITATION GOALS

The goals of rehabilitation depend on the effects of the illness, what the patient was able to do before the illness, and the patient's wishes. Establishing goals of rehabilitation helps determine the setting and method of rehabilitation. Wherever possible the goal of rehabilitation is to achieve full, unrestricted function. In most of the patients, the goal of rehabilitation is often limited to restoration of the ability to perform as many activities of daily living (ADLs) as possible.

Timing

As soon as patients are medically stable rehabilitation process can be started. This will help to prevent secondary disabilities (e.g., contractures) and depression.

Role of Family

Family education is an important part of the discharge process, particularly when the patient is discharged into the community. Family members are taught how to help the patient be as independent as possible, so that they do not overprotect the patient (leading to decreased functional status and increased dependence) or neglect the patient's primary needs (leading to feelings of rejection, which may cause depression or interfere with physical functioning).

Special Equipments

Even after rehabilitation, some will need support for walking, balancing, or performing certain activities of daily living. Special equipment can sometimes help (Box)

- Cane
- Walker
- Ankle-foot orthotic devices (braces)
- Wheelchair
- Aids for bathing, dressing, and eating
- Communication aids

Box 49.3 Key points in success

Patient's general condition
Range of motion
Muscle strength
Bowel and bladder function
Premorbid functional and cognitive ability
Social situation
Learning ability
Motivation
Coping skills
Ability to participate in rehabilitation

Continuum of Care

Medical rehabilitation rarely is conducted in only one care setting. This continuum of care has become increasingly complex, ranging from high-cost and high-intensity care in acute care hospitals, to lower-cost and lower-intensity care in outpatient settings. Assessment of patients across the continuum of care should be understandable, meaningful, cost-effective, and manageable.

Discharge Planning

This should begin immediately on admission to hospital. The patient and his or her relatives should be made aware that discharge is anticipated, and the expectation either of prolonged or even permanent hospitalization must be prevented. When the patient is fit to get out of bed, the natural history of the illness and the time course of recovery require consideration, and the presence of complications should be assessed, including in particular pressure lesions of the skin and postural hypotension. Prevention of the former is an important immediate aspect of rehabilitation, since a full-thickness pressure sore will retard rehabilitation by many weeks; its creation takes a few hours. Postural hypotension may be due to drugs, to biochemical disorders, or to bed rest itself. The patient's mental state requires assessment, with particular attention to intellectual impairment, the presence of depression, and the adequacy of morale. Disability must be assessed in functional terms (for example the ability to feed, dress, maintain continence, and walk) and must be seen against the state preceding the acute illness. A target date for discharge should be set, and arrangements made with relatives and community services to ensure that support is available on the day of discharge and immediately thereafter. When the drug regimen has been reduced to the minimum necessary, it should be explained in detail to the patient, who should be given the responsibility for it, supervised by the nursing staff or ward pharmacist.

METHODS OF REHABILITATION

The physician or rehabilitation team determines which methods of rehabilitation are appropriate.
Physical therapy
Leisure or recreational therapy
Occupational therapy

Physical Therapy

Before prescribing physical therapy, one should ensure that the patient is medically stable and notes any cardiac, pulmonary, neurologic, or musculoskeletal limitations.

Range-of-motion Exercises (Table 49.1)

Several physical therapy techniques can help improve range of motion, which commonly becomes restricted after a stroke or prolonged bed rest. Restricted range of motion can cause pain, reduce functional abilities, and predispose patients to pressure sores. Range of motion should be evaluated with a goniometer before therapy and regularly thereafter.

One should remeber that in elderly persons, the range of motion for certain joints is usually lower than would be normal for younger patients.

Table 49.1 Examples of range-of-motion exercises

Active	For patients who can exercise without assistance
Active assistive	For patients whose muscles are too weak to exercise without assistance. For patients who experience discomfort during joint moyement
Passive	For patients who cannot actively participate

Patient Evaluation

Before beginning therapy, the physical therapist must determine if restricted motion is due to tight ligaments and tendons or to tight muscles.
Active-assistive or passive range-of-motion exercises must be performed very gently as aggressive movements can easily damage joints with restricted motion or break osteoporotic bones.
The affected joint must be moved beyond the point of pain, but the movement should not cause residual pain.
Movements producing severe pain should be avoided, although some discomfort may be unavoidable.
If the affected joint is adjacent to an unfixed fracture, passive exercises should not be performed.
Sustained moderate stretching is more effective than momentary forceful stretching.

Muscle-strengthening Exercises

The purpose of these exercises is to strengthen muscles enough to perform a given function, not necessarily to regain normal strength for age.
When a muscle is very weak, gravity alone is sufficient.
Many forms of exercise increase muscle strength; all involve progressively increased resistance.
As muscle strength increases, resistance is gradually increased.

Proprioceptive Neuromuscular Facilitation

This technique promotes useful neuromuscular activity in patients with spasticity due to upper motor neuron damage; it enables them to feel muscle contraction and helps maintain the affected joint's range of motion. For example, in patients with right hemiplegia, strong resistance applied to the left biceps causes the right elbow to flex through contraction of the right (hemiplegic) biceps. The exact mechanism is not clearly understood, but reflex-related proprioception may be involved. Various techniques (e.g., Brunnstrom, Rood, Bobath) are widely used.

Coordination Exercise

These task-oriented exercises are for patients who need to improve coordination (e.g., stroke patients). They involve repeating a meaningful movement that works more than one joint and muscle (e.g., picking up an object, touching a body part). The goal is to improve motor skills to the premorbid level.

Transfer Training

Patients who cannot transfer safely have a high risk of falling, with risk of fracture or other injury. The techniques used depend on whether the patient can bear weight on one or both legs, has sound balance, or has hemiplegia. Assistive devices can sometimes help. For example, persons who have difficulty standing from a seated position may benefit from a chair with a raised seat or a self-lifting chair.

Sitting

Patients can safely sit up once they are fully conscious and their neurologic deficits are no longer progressing.

Ambulation Exercises

The purpose of these exercises is to improve the patient's ability to walk independently or be assisted by a person or device. The goal of ambulation exercises is to establish and maintain a safe gait, not to restore a normal gait. Before ambulation exercises can be started, patients must be able to stand. Patients first learn to stand from the sitting position. Patients must stand with the hips and knees fully extended, leaning slightly forward and towards the unaffected side. Using the parallel bars is the safest way to practice standing.

Before starting ambulation exercises, some patients need to improve a joint's range of motion or muscle strength. If a muscle remains weak or spastic, an orthotic device (e.g., a brace) may be necessary. Training may begin on parallel bars, especially if the patient's balance is impaired, and progress to walking with aids (e.g., walker, crutches, cane).

Use of a Tilt Table

For patients with orthostatic hypotension due to paraplegia, quadriplegia, prolonged bed rest, or immobilization, a tilt table may be used to help reestablish hemodynamic balance. The patient, held in place with a safety belt, lies supine on a padded table with a footboard. The table is tilted manually or electrically; the angle is increased very slowly to 85°, if this angle can be tolerated. How long the position is maintained depends initially on the patient's continued tolerance, but it should not exceed 45 minutes.

Stair Climbing

For stair climbing, ascent starts with the better leg, and descent with the affected leg (good leads up; bad leads down). If possible, patients ascend and descend with the railing on the unaffected side, so that they can grasp the railing. During descent, the patient should use a cane. The cane should be moved to the lower step shortly before descending with the bad leg.

General Conditioning Exercises

A combination of the exercises described above is used to counter the effects of debilitation, prolonged bed rest, or immobilization; to reestablish hemodynamic balance; to increase cardiorespiratory capacity; and to maintain range of motion and muscle strength.

TREATMENT OF PAIN AND INFLAMMATION

Heat Therapy

Mechanism of Action

Heat increases blood flow and the extensibility of connective tissue
Decreases joint stiffness, pain, and muscle spasm
Reduces inflammation, oedema, and exudates resolve
The intensity and duration of heat's physiologic effects are determined mainly by tissue temperature, the rate of temperature elevation, and the area treated.

Indications

Acute and chronic traumatic and inflammatory conditions (e.g., sprains, strains, fibrositis, tenosynovitis, muscle spasm, myositis, painful back, whiplash injuries, various forms of arthritis, arthralgia, neuralgia). Heat must be applied very carefully to patients because skin sensation or cognitive capacity may be diminished, increasing the risk of burns.

Types of applications

Superficial	Hot packs Infrared heat Paraffin baths Hydrotherapy
Deep	Diathermy Ultrasound

Table 49.2 Methods of applications of heat therapy

Type	Method	Indications	Contraindications
Hotpacks	Cotton cloth containers filled with silicate gel	Boiled in water, cooled to a temperature that does not burn the skin, and applied Wrapping the packs in several layers of towels helps protect against burns	Advanced heart disease Peripheral vascular disease Impaired skin sensation (particularly to temperature and pain)
Infrared	Applied with a lamp	Usually for 20 minutes/day	Same as above
Paraffin bath	Wax heated to 49°C (120°F)-never > 54.4°C (> 130°F)	Used to apply heat, usually to small joints Applied by dipping or immersing (e.g., a hand) or painting (e.g., a knee or elbow) and then wrapping with a towel	Open wounds Contraindicated in persons allergic to paraffin
Hydrotherapy	Agitated warm water 35.5° to 37.7°C (96° to 100°F)	Relax muscles and relieve pain Particularly useful in conjunction with range-of-motion exercises	Same as hotpacks

contd.

Short wave diathermy	Less effective than previously thought	Used to treat inflammation	Malignancy Haemorrhagic conditions Peripheral vascular disease Loss of sensation Persons with nonremovable prostheses Electrophysiologic braces Metallic implants
Microwave diathermy	More comfortable to apply than short wave diathermy. Output measurement is more accurate	Used to treat pain and inflammation	Same as short wave diathermy

Ultrasound

Ultrasound involves the use of high-frequency sound waves, which penetrate deep into the tissue (4 to 10 cm [1.6 to 4 inches]) and produce thermal, mechanical, chemical, and biologic effects. This therapy may be used to treat limited range of motion caused by muscle shortening and fibrosis; skin or subcutaneous tissue scarring; bursitis, calcific bursitis, tendinitis, myositis, tenosynovitis, epicondylitis, and spondylitis; pain from postoperative neurofibromas (especially those embedded in scar tissue); myofascial pain syndrome, phantom pain, neuritis; sciatica and other forms of radiculitis; contusions; reflex dystrophies (e.g., Sudeck's atrophy, causalgia, shoulder-hand syndrome); and chronic skin ulceration. Ultrasound is contraindicated in patients with ischaemic tissue, haemorrhagic diathesis, malignancies, anaesthetized areas, or areas of acute infection. Also, it should not be applied over the eyes, brain, spinal cord, ears, heart, reproductive organs, brachial plexus, or healing bone.

Cold Therapy (cryotherapy)

Application of cold may help relieve muscle spasm, myofascial or traumatic pain, acute low back pain, and acute inflammatory lesions as well as help induce local anaesthesia. The choice between heat or cold therapy is often empiric; however, for acute pain, cold therapy seems to be more effective than heat therapy. Cold may be applied locally using an ice bag, a cold pack, or volatile fluids (e.g., ethyl chloride), which cool by evaporation.

Electrical Stimulation

Denervated skeletal muscle and innervated muscle that cannot be contracted voluntarily can be stimulated electrically to help alleviate or prevent disuse atrophy and muscle spasticity, especially in patients with hemiplegia due to a cerebrovascular accident, with traumatic paraplegia or quadriplegia, or with peripheral nerve injury. Electrical stimulation is contraindicated in patients with advanced cardiac disease, because it may precipitate an arrhythmia, and in patients with a pacemaker, because it may interfere with its functioning. Electrical stimulation should not be applied over the eyes.

Transcutaneous Electrical Nerve Stimulation (TENS)

Transcutaneous electrical nerve stimulation (TENS), which uses low current at low-frequency oscillation, is particularly useful for chronic back pain, rheumatoid arthritis, sprained ankle, contusion, postherpetic neuralgia, causalgia, phantom limb syndrome, and trigger points. It may also promote callus formation in a nonunited fracture. TENS may be applied several times daily for 20 minutes to several hours, depending on the severity of pain. The device produces a gentle tingling sensation without increasing muscle tension. TENS is contraindicated in persons with advanced cardiac disease or a pacemaker because it may precipitate an arrhythmia. It should not be applied over the eyes.

Traction

Spinal traction is used to overcome extrinsic muscle spasm and to keep bony surfaces aligned while fractures heal. A weight and pulley system, the patient's weight, or manual or motorized force can be used. The force may be applied continuously or intermittently.

Cervical traction is often used for chronic neck pain due to cervical spondylosis, disk prolapse, whiplash, or torticollis. A 5–10 lb (2.5–5 kg) weight is used. Some advocate heavier weights, but sustained traction with > 20 lb (> 10 kg) for more than a few minutes is poorly tolerated; motorized intermittent rhythmic traction is generally well tolerated. Generally, hyperextension of the neck should be avoided, because it may increase root compression in the neuroforamina.

Lumbar traction is rarely used, although it is sometimes recommended for treating patients with painful lumbar osteoarthritis or spondylolisthesis. Its value in treating acute discogenic pain is debated, and it puts the elderly at risk of developing a secondary disability because it requires prolonged bed rest. For patients with severe osteoporosis or osteoarthritis, traction must be carefully and gently applied.

Massage

Massage may relieve pain, reduce swelling and induration due to trauma (e.g., fracture, joint injury, sprain, strain, bruise, peripheral nerve injury), and mobilize contracted tissues. Massage may be appropriate for patients with low back pain, arthritis, periarthritis, bursitis, neuritis, fibrositis, hemiplegia, paraplegia, quadriplegia, multiple sclerosis, or cerebral palsy. It should not be used to manage infections or thrombophlebitis.

Acupuncture

Thin needles are inserted through the skin at specific body sites, frequently far from the site of pain. These needles, made of stainless steel, gold, or platinum, are twirled rapidly and intermittently for a few minutes, or a low electric current is applied through the needles. Although the mechanism of action is not fully understood, many practitioners believe that acupuncture stimulates endorphin production, generating analgesic and anti-inflammatory effects. The value of this technique is debated. Acupuncture should be performed only by trained persons. Sterilized or new needles must be used to avoid infection.

BIOFEEDBACK

Introduction

Biofeedback operates on the notion that a person has the innate ability and potential to influence the automatic functions of his own body through the exertion of will and mind.

Historical Perspective

The word "biofeedback" was coined in the late 1960s to describe laboratory procedures then being used to train experimental research subjects to alter brain activity, blood pressure, heart rate, and other bodily functions that normally are not controlled voluntarily. At the time, many scientists looked forward to the day when biofeedback would give a major degree of control over body functions. Research has demonstrated that biofeedback can help in the treatment of many diseases and painful conditions.

Mechanism of Action

Scientists cannot yet explain how biofeedback works but most patients who benefit from biofeedback are able to relax and modify their behaviour. Stressful events produce strong emotions, which arouse certain physical responses. Many of these responses are controlled by the sympathetic nervous system, that helps prepare the body to meet emergencies by "flight or fight." Relief of this stress is the key component in the success of biofeedback treatment modalities.

Clinical Applications of Biofeedback

Clinical biofeedback techniques that grew out of the early laboratory procedures are now widely used to treat an ever-lengthening list of conditions. These include:

- Migraine headaches, tension headaches
- Management of chronic back pain
- Disorders of the digestive system (irritable bowel syndrome, constipation)
- High blood pressure
- Low blood pressure
- Cardiac arrhythmias
- Raynaud's disease
- Epilepsy
- Paralysis and other movement disorders (rehabilitation following a stroke)

Biofeedback Techniques

Several different types of biofeedback machines can provide information about the systems in your body that are affected by stress.

Electromyogram (EMG)

The electromyogram (EMG) measures muscle tension. The most common muscles that are used for biofeedback are the frontalis, the masseter, and the trapezius. The EMG has been utilized to rehabilitate patients paralyzed by stroke. In these patients EMG can detect even small electrical activity in the paralyzed muscles and as the patient becomes aware of the activity, his nervous system may stimulate more muscle activity. This will help more new nerve endings to grow in the affected muscles and to regain some motor activity in stroke patients. EMG had also been used for the treatment of tension headaches, backache, neck pain, and bruxism as well as in the stress related illnesses such as asthma and ulcers.

Temperature Biofeedback

Temperature biofeedback device monitors skin temperature and can be helpful in certain circulatory disorders. Reynolds disease is an example that can be benefited by this technique. Usually, a sensor is attached to the foot or to the middle or small finger of dominant hand. It is based on the assumption that when a person is tense or anxious, his skin temperature drops as blood is redirected inward to muscles and internal organs.

Like monitoring muscle tension, measuring skin temperature is a useful tool in learning how to manage stress. This method may also reduce the frequency of migraine headaches, and is also used to promote relaxation.

Electroencephalogram (EEG)

On EEG alpha waves are commonly observed when a person relaxes. It is presumed that patients can relief from anxiety, insomnia and perhaps epilepsy by learning to increase their alpha wave activity. This alpha training is only useful if it is combined with other therapies.

Galvanic Skin Response (GSH)

Also known as electrodermal response (EDR), Galvanic skin response measures electrical conductance in the skin, which is associated with the activity of the sweat glands. A very slight electrical current (unnoticeable to you) is run through the patient's skin. Then a machine measures changes in the salt and water in the person's sweat gland ducts. The more emotionally aroused the patient is, the more active will be the sweat glands, and the greater the electrical conductivity of the skin. GSR is effective in treating phobias, anxiety, excessive sweating, and, stuttering.

Conclusion

Biofeedback is a tool that can be used for health care, but it cannot cure disease or by itself make a person healthy.

CHAPTER 50

Disability Evaluation and Management

DISABILITY EVALUATION

"Handicap" and "disability" are often used as interchangeable terms, referring generally to the problems arising from disease. The WHO definitions serve to increase precision of thought, and to improve communication.

Impairment

"Any loss or abnormality of psychological, physiological or anatomical structure or function", i.e., systems or parts of the body that do not work.

Disability

"Any restriction or lack (resulting from an impairment) of ability to perform an activity in the manner, or within the range, considered normal for a human being" i.e., things people cannot do.

Handicap

"A disadvantage for a given individual, resulting from an impairment or disability, that limits or prevents the fulfillment of a role (depending on age, sex, and social or cultural factors) for that individual" i.e., social consequences of impairments and/or disabilities.

ASSESSING IMPAIRMENTS (TABLE 50.1 TO TABLE 50.10)

Impairments can be assessed using the whole range of clinical method, from bedside estimates, through simple measurements, to complex high-tech applications. Many impairments are assessed routinely as symptoms and signs (e.g., limb weakness may be graded using the MRC muscle power scale from grade 0–V).

Table 50.1 Spinal cord levels and extent of disabilties

Spinal cord levels	C1 to C3
Key muscles innervated	Sternocleidomastoid Levator scapulae Upper trapezius Diaphragm (C3–C5)
Movements possible	Neck control Chew Swallow Talk Sip Puff Some capular elevation
Pattern of weakness	Complete paralysis of trunk, upper extremities (UEs), and lower extremities (LEs) Dependence on respirator
Functional potential	Requires full-time attendant care Total dependence with activities of daily living (ADLs) and transfers Can propel power wheelchair equipped with portable respirator and chin, head, puff, or sip controls Can operate communication and environmental control systems with head master, head pointer, mouth stick, or pneumatic control

Table 50.2

Spinal cord levels	C4
Key muscles innervated	Trapezius (superior, middle, and inferior) Diaphragm Cervical and paraspinal muscles
Movements possible	Respiration Scapula elevation Neck movements
Pattern of weakness	Paralysis of trunk UEs and LEs (except scapula elevation)

contd.

Functional potential	Good potential to control breathing without ventilator Requires full-time attendant care Can drink with long straw Total dependence with ADLs and transfers Can independently power wheelchair with chin, head, sip, or puff controls Activities can be accomplished through use of mouth stick, head pointer, voice recognition software, or tongue touch key pad

Table 50.3

Spinal cord levels	C5
Key muscles innervated	Partial deltoid Biceps brachii Brachialis Brachioradialis Rotator cuff muscles rotation Rhomboids Serratus anterior Teres major All shoulder muscles are at least partially innervated except coracobrachialis latis simus dorsi
Movements possible	Shoulder abduction, flexion, extension, horizontal abduction, horizontal adduction, internal and external rotation Scapular protraction and retraction Elbow flexion and supination
Pattern of weakness	Total paralysis of trunk and LEs Low endurance because of paralysis of intercostals and low respiratory reserve No active elbow extension, forearm pronation, hand or wrist movement
Functional potential	With adaptive equipment or splints and set-up assistance, can perform eating, handwriting, light hygiene, shaving, telephoning and typing May be independent in upper body dressing, if muscle strength is good Otherwise may need minimum to moderate assistance Dependent in lower body with transfers, may be able to assist in rolling in bed, using side rails, or loops, independent with power wheelchair May drive a van with substantial adaptations Can write independently with wrist support and writing device Requires a minimum of at least part-time attendant care

Table 50.4

Spinal cord levels	C6
Key muscles innervated	Extensor carpi radialis longus and brevis Serratus anterior (partial but significant innervation) Pronator teres Coracobrachialis Pectoralis major Latissimus dorsi
Movements possible	Full strength with shoulder movements (flexion, extension, abduction, adduction, external, and internal rotation) Forearm pronation and supination Radial wrist extension Gross grasp and gross prehension via tenodesis action Good stability of scapular on trunk
Pattern of weakness	No elbow extension or ulnar wrist extension Endurance may be low No active wrist flexion Complete paralysis of trunk and lower extremities
Functional potential	Able to perform many activities independently with tenodesis splint or universal cuff, such as self-feeding with regular utensils, personal hygiene, and grooming Independent in upper body dressing Minimal assistance for lower body dressing Bed mobility, independence in rolling side-to-side Minimal assistance from supine to sitting Assist with transfers by substituting shoulder adduction and external rotation for elbow extension May be independent with a sliding board Independent with manual wheelchair on level terrain or slight incline Requires adaptive equipment and assistance for bathing and bowel care Independent at driving car with hand controls May participate in wheelchair sports May require part-time attendent care

Table 50.5

Spinal cord levels	C7
Key muscles innervated	Triceps brachii Pectoralis major Latissimus dorsi Flexor digitorum superficialis Extensor digitorum Flexor carpi radialis Extensor carpi ulnaris Extensor pollicis longus and brevis Abductor pollicis longus
Movements possible	Elbow extension Full strength of all shoulder movements Finger flexion but weak Finger MP joint extension Radial wrist flexion Ulnar wrist extension Thumb extension
Pattern of weakness	Limited grasp, release, and dexterity Complete paralysis of trunk and lower extremities
Functional potential	Independent in ADLs Can perform transfers independently Independent bed mobility Independent for push-up in wheelchair for pressure release Independent with driving with hand controls Independent with bowel and bladder care Independent with manual wheelchair

Table 50.6

Spinal cord levels	C8 to T1
Key muscles innervated	All the muscles of the UE are now fully innervated Dorsal and palmar interossei Lumbricals Thenar and hypothenar muscles Adductor pollicis Flexor digitorum profundus Flexor pollicis longus and brevis Pronator quadratus
Movements possible	Full control of upper extremities, including finger flexion, isolated finger and thumb movements, fine coordination, and grasp
Pattern of weakness	Paralysis of lower extremities Weakness of trunk control Lower respiratory reserves
Functional potential	Independent in ADLs, with transfers and bed mobility

Table 50.7

Spinal cord levels	T4 to T9
Key muscles innervated	All muscles of upper extremity Partial innervation of intercostal muscles and long muscles of the back
Movements possible	Full arm function Partial trunk stability Increased endurance
Pattern of weakness	Total paralysis of lower extremities Partial trunk paralysis
Functional potential	Independent in all self care, independent manual wheelchair use, and transfers Drives car independently with adaptations May use standing frame independently Independent light housekeeping

Table 50.8

Spinal cord levels	T10 to L2
Key muscles innervated	Intercostal muscles fully innervated Abdominal muscles are partially to fully innervated
Movements possible	Good trunk stability Increased physical endurance
Pattern of weakness	Paralysis of lower extremities
Functional potential	Independent in self care, work, personal hygiene, and housekeeping Drives car with hand controls Often uses wheelchair but can ambulate with difficulty using crutches and braces

Table 50.9

Spinal cord levels	L3 to L4
Key muscles innervated	Lower back muscles Quadriceps Hip flexors Hip adductors
Movements possible	Trunk stability and control Hip flexion Hip adduction Knee extension
Pattern of weakness	Individual cannot perform hip extension, knee flexion, or ankle and foot movements May have weakness of ankle and foot
Functional potential	Can ambulate independently with short leg braces using crutches May still use a wheel chair for energy conservation

Table 50.10

Spinal cord levels	L5 to S3
Key muscles innervated	Gluteus maximus Hamstrings Knee flexors Ankle and foot muscles
Movements possible	Partial to full control of lower extremities
Pattern of weakness	Possible decrease in standing and walking tolerance modifications
Functional potential	Independent in all activities Can drive car without modification May require braces for ambulation

SENSATION ASSESSMENTS

Surface Sensation

Proprioceptive Sense

Functional Assessments

Speed and coordination test
Hand reaction
Balancing
Gait evaluation
Facial movements

MUSCLE TONE ASSESSMENT

Reflexes

Deep tendon reflex
Superficial reflexes

Deep tendon reflex grading (Common in the biceps, triceps, quadriceps, and Achilles tendon)

0	No response
+	Hypoactive response
++	Normal response
+++	Hyperactive response
++++	Hyperactive response

The modified Ashworth scale

0	No increase in muscle tone
1	Slight increase in tone, minimal resistance at the end of the ROM
1+	Slight increase in tone, minimal resistance throughout the remainder (less than half) of the ROM
2	More remarked increase in muscle tone through the most of ROM
3	Considerable increase in muscle tone, passive movement difficult
4	Affected part (s) rigid in in flexion or extension

ASSESSMENT OF DISABILITY

Frequently, details of the impairment do not allow accurate prediction of the extent of disability as similar limb weakness (impairment) may produce different degrees of disability. There is a vast range of different disabilities, but of prime importance is the degree of independence in the activities of daily living (ADL), which cover basic self-care and mobility. There are many different scales for assessing disability.

- Barthel ADL index
- Modified Barthel score
- FIM (The functional independence measure)
- Rivermead mobility index
- SF–36
- 12-item health status questionnaire (HSQ–12)

The Barthel Index (Table 50.11)

The Barthel index records that disability is present, and estimates its extent It does not take account of underlying causes nor indicates potential for improvement. The Barthel index is used fairly universally in hospitals for assessing patients with severe disabilities.

Table 50.11

1. Bowel status	0 points – Incontinent (or needs to be given enema) 1 point – Occasional accident (once a week) 2 points – Fully continent
2. Bladder status	0 points – Incontinent or catheterized and unable to manage 1 point – Occasional accident (max once per 24 hours 2 points – Continent (for more than seven days)
3. Grooming	0 points – Needs help with personal care: face/ hair/teeth/shaving 1 point – Independent (implements provided)
4. Toilet use	0 points – Dependent 1 point – Needs some help but can do somethings alone 2 points – Independent (on and off/ wiping/ dressing)
5. Feeding	0 points – Unable 1 point – Needs help in cutting/spreading butter/etc. 2 points – Independent (food provided within reach)
6. Transfer	0 points – Unable (as no sitting balance) 1 point – Major help (physical/one or two people) 2 points – Can sit minor help (verbal or physical) 3 points – Independent
7. Mobility	0 points – Immobile 1 point – Wheelchair-independent (including corners/etc.) 2 points – Walks with help of one person (verbal or physical) 3 points – Independent
8. Dressing	0 points – Dependent 1 point – Needs help but can do about half unaided 2 points – Independent (including buttons/zips/laces/etc.)
9. Stairs	0 points – Unable 1 point – Needs help (verbal/physical/carrying aid) 2 points – Independent up and down
10. Bathing	0 points – Dependent 1 point – Independent bathing or showering

Barthel Score (max 20)

Modified Barthel Score (Table 50.12)

This modification further increases the sensitivity of the score (maximum 100), without increasing difficulty undertaking test or time involved

Table 50.12

Items	Unable to perform task	Attempts task but unsafe	Moderate help required	Minimal help required	Fully independent
Personal hygiene	0	1	3	4	5
Bathing self	0	1	3	4	5
Feeding	0	2	5	8	10
Toilet	0	2	5	8	10
Stair climbing	0	2	5	8	10
Dressing	0	2	5	8	10
Bowel control	0	2	5	8	10
Bladder control	0	2	5	8	10
Ambulation (wheelchair)	0 (0)	3 (1)	8 (3)	12 (4)	15 (5)
Chair-bed transfers	0	3	8	12	15

Total = 100

The Functional Independence Measure

A basic indicator of the severity of disability and the burden of care 7-point scale

With the FIM and other standardized assessment tools, the therapist is able to:

- Make objective measures of functional mobility
- Enhance communication of a patient's status and progress within and across disciplines
- Provide uniformity of documented outcomes
- Measure treatment efficacy

CHAPTER 51

Activities of Daily Living

The activities of daily living (ADLs) are a defined set of activities necessary for normal self-care. The activities are movement in bed, transfers, locomotion, dressing personal hygiene, and feeding (Table 51.1).

Table 51.1 Activities of daily living

Movement in bed	Means sitting in, rising from, and moving around in bed
Transfers	Means moving from one seat to another, changing position from sitting to standing, and transferring to and from the toilet and bed
Locomotion	Means walking on the level, on gentle slopes and down stairs
Dressing	Means putting on socks, stockings, and shoes, as well as clothing the upper and lower trunk
Personal hygiene	Means grooming, and washing of face, trunk, extremities and perineum
Feeding	Means eating and drinking, but not the preparation of food

REHABILITATION ASPECT OF ADL

Role of the physiotherapist is to assist patients with difficulties in the performance of activities of daily living, including self-feeding, dressing, grooming, bathing and toileting, as well as iob performance.

Various pieces of adaptive equipment may be recommended to increase success and independence with activities of daily living. The specific components for these systems should always be determined by a rehabilitation team that has experience with adaptive equipments. Some specific examples include:

Self-feeding

Built-up handles to improve grasp on an object.
Bent-angled spoons to decrease wrist turning as food nears the mouth
Whole-handed grasp of a spoon by the shaft of the handle to increase stability.
Elbows stabilized on the table to steady the hand.
Decreasing the distance of hand to mouth movement by placing a 6 or 12-inch platform under the plate to help decrease spillage.
Choosing finger foods and textures such as mashed potatoes that are easy to self-feed.

Bathing

Tub bars to increase safety and independence with performance of bathing.
Bath and shower chair to decrease fatigue, increase safety and increase the ability to bathe independently.
A portable showerhead with a bath seat to shower in the bathtub.

Toilet

A raised toilet seat makes coming to a standing position more easy.
Toilet rails to increase safety and independence with performance of toileting.
Pants can be modified (e.g., Velcro flap replaces a zipper)

Dressing

Modify clothing to simplify dressing, undressing and arranging clothes for toileting (i.e., loose fitting clothing with elastic waist bands, without fasteners, and modified shoe closures (self-holding shoe strings, Velcro closure). Dressing in a chair or sitting against a wall or the base of a bed may be useful to improve trunk stability and "free up" the arms to assist with the task.

Home Environment and Safety

Wider door frames for easy wheelchair access throughout the home, along with tiled floors for easier wheelchair movement.
Modification of kitchen table and/or computer table to allow the wheelchair to be used.
Where possible, sit rather than stand.
Work in front of rather than at the side.
Sliding rather than lifting objects.
Try to maintain good posture when standing, bending or sitting
Avoid loose or floppy slippers or shoes.

CHAPTER 52

Brain Death and Persistent Vegetative State

BRAIN DEATH

Definition

Brain death is defined as an irreversible and complete loss of function of the entire brain including the brain stem. Death is a process and not an event, in that organs cease to function in several sequences.

Causes of Brain Death

Head injury
Spontaneous intracranial haemorrhage
Brain tumour or infection

Clinical Examination

The clinical examination of patients who are presumed to be brain dead must be performed with precision. The declaration of brain death requires not only a series of careful neurologic tests but also the establishment of the cause of coma, the ascertainment of irreversibility, the resolution of any misleading clinical neurologic signs, the recognition of possible confounding factors, the interpretation of the findings on neuroimaging, and the performance of any confirmatory laboratory tests that are deemed necessary.

Neurologic Examination

Neurologic examination to determine whether a patient is brain dead can proceed only if the following prerequisites are met: the ruling out of complicated medical conditions that may confound the clinical assessment, particularly severe electrolyte, acid–base, or endocrine disturbances; the absence of severe hypothermia, defined as a core temperature of 32°C or

lower; hypotension; and the absence of evidence of drug intoxication, poisoning, or neuromuscular blocking agents.

Complete clinical neurologic examination includes documentation of coma, the absence of brain-stem reflexes, and apnea. The examination of brain-stem reflexes requires the measurement of reflex pathways in the mesencephalon, pons, and medulla oblongata. As brain death occurs, patients lose their reflexes in a rostral-to-caudal direction, and the medulla oblongata is the last part of the brain to cease to function. Several hours may be required for the destruction of the brain stem to be complete, and during that period, there may still be medullary function. An obligatory prerequisite for the diagnosis of brain death is the proof of severe primary or secondary brain damage.

Box 52.1 Clinical criteria for brain stem death in adults and children

Coma
Absence of motor response
Absence of pupillary response to light and pupils at mid-position with respect to dilatation (4–6 mm)
Absence of corneal reflex
Absence of gag reflex
Absence of coughing in response to tracheal suctioning
Absence of sucking and rooting reflexes
Absence of respiratory drive at a $PaCO_2$ that is 60 mm Hg or 20 mm Hg above normal baseline values
Interval between two evaluations, according to patient's age
- Term to 2 months old, 48 h
- > 2 months to 1 year old, 24 h
- > 1 year to 18 year old, 12 h
- > 18 year old, interval optional

Confirmatory tests (Box)
- Term to 2 months old, 2 confirmatory tests
- > 2 months to 1 year old, 1 confirmatory test
- > 1 year to 18 years old, optional
- > 18 years old, optional

Box 52.2 Confirmatory testing for a determination of brain death

Cerebral angiography
Electroencephalography
Transcranial Doppler ultrasonography
Cerebral scintigraphy (technetium Tc 99m hexametazime)

Steps of Examination for Brain Death

Step 1

The physician determines that there is no motor response and the eyes do not open when a painful stimulus is applied to the supraorbital nerve or nail bed.

Step 2

A clinical assessment of brain-stem reflexes is undertaken.
The absence of grimacing or eye opening with deep pressure on both condyles at the level of the temporo-mandibular joint (afferent nerve V and efferent nerve VII)
The absent corneal reflex elicited by touching the edge of the cornea (V and VII)
The absent light reflex (II and III)
The absent oculo-vestibular response toward the side of the cold stimulus provided by ice water (pen marks at the level of the pupils can be used as reference VIII and III and VI)
The absent cough reflex elicited through the introduction of a suction catheter deep in the trachea (IX and X).

Step 3

The apnoea test is performed; the disconnection of the ventilator and the use of apnoeic diffusion oxygenation require precautionary measures. The core temperature should be 36.5°C or higher, the systolic blood pressure should be 90 mm Hg or higher, and the fluid balance should be positive for six hours. After preoxygenation (the fraction of inspired oxygen should be 1.0 for 10 minutes), the ventilation rate should be decreased. The ventilator should be disconnected if the partial pressure of arterial oxygen reaches 200 mm Hg or higher and if the partial pressure of arterial carbon dioxide reaches 40 mm Hg or higher. The oxygen catheter should be at the carina (delivering oxygen at a rate of 6 liters per minute). The physician should observe the chest and the abdominal wall for respiration for 8 to 10 minutes and should monitor the patient for changes in vital functions. If there is a partial pressure of arterial carbon dioxide of 60 mm Hg or higher or an increase of more than 20 mm Hg from the normal base-line value, apnoea is confirmed.

Implications

After the clinical criteria of brain death have been met, the physician should inform the next of kin, who can be approached about organ donation. The physician is required to abide by state law with respect to organ donation. Organ-procurement agencies must be notified to request the donation of organs. If the legal next of kin declines to donate organs, it is good medical

judgment to discontinue mechanical ventilation. When mechanical ventilation and support are continued because of ethical or legal objections to their discontinuation, what usually follows is an invariant heart rate from a differentiated sinoatrial node, structural myocardial lesions leading to a marked reduction in the ejection fraction, decreased coronary perfusion, the need for increasing use of inotropic drugs to maintain blood pressure, and a fragile state that leads to cardiac arrest within days or weeks.

THE PERSISTENT VEGETATIVE STATE

The persistent vegetative state is defined as the clinical condition resulting from loss of function in the cerebral cortex with a functioning brain-stem. Because of intact brain stem function, vegetative patients breathe spontaneously and are not ventilator-dependent; and can survive for many years, if adequately fed and nursed.

Box 52.3 Common causes of persistent vegetative state

- Severe head injury
- Severe diffuse axonal injury
- Secondary hypoxic brain damage to the brain following
  - Cardiac arrest
  - Near drowning
  - Strangulation
- Severe hypoglycaemia
- Intracranial infections causing asphyxia in children
- End-stage of Alzheimer's disease
- End-stage of multi-infarct dementia
- Devastating congenital cerebral defects

Diagnosis

Diagnosis depends on characteristic clinical features recorded by skilled observers over a period of time.

Box 52.4 Characteristics of persistent vegetative state

- The patient has long periods of spontaneous eye opening.
- The eyes may briefly follow a moving object and the head turn reflexly to a sudden noise that produces a startle reaction.
- All four limbs are paralysed and spastic, with only reflex posturing and withdrawal.
- The face may grimace and groans be heard, but never words.
- There is no psychologically meaningful response to external stimuli or any learned behaviour.
- No evidence of a working mind.

These patients need to be distinguished from patients with a locked in syndrome who are aware and can communicate by the only remaining motor power, namely by blinking or eye movements.

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